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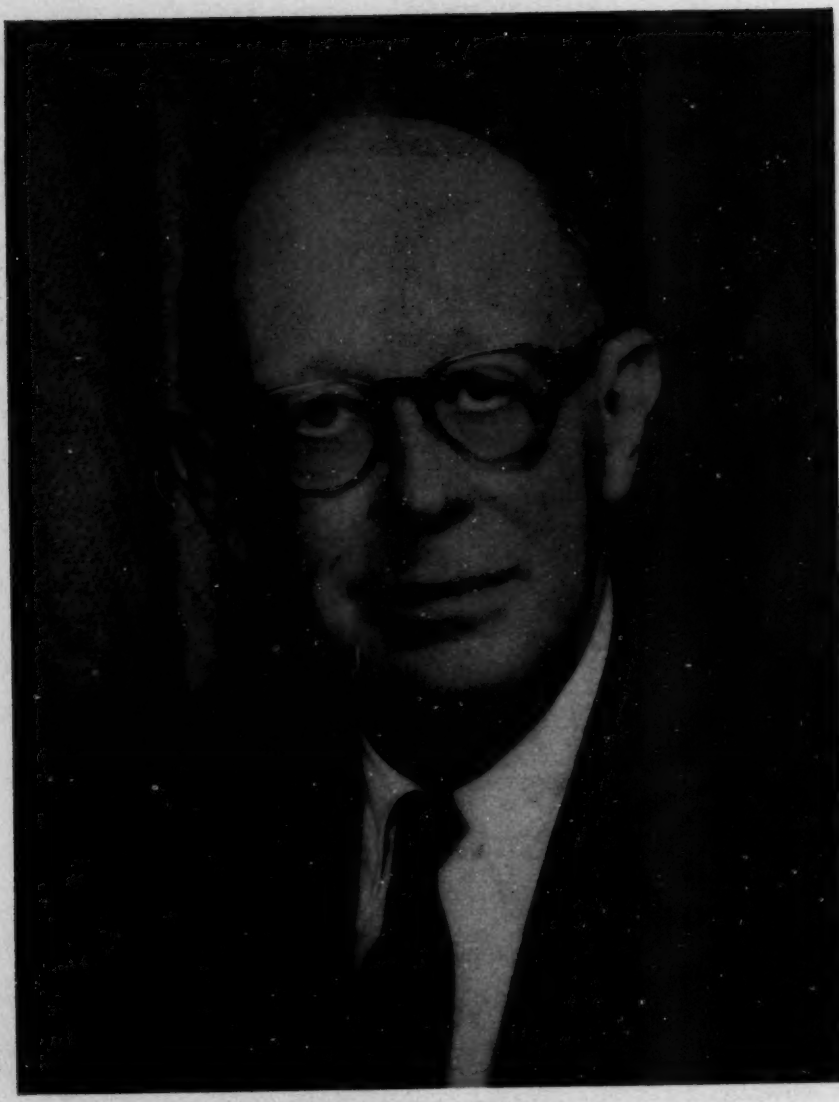

Winthrop M. Phelps

PERSONALITIES OF TODAY

Winthrop M. Phelps, M.D.

By JOHN H. CROSLAND

WINTHROP PHELPS was born in the township of Bound Brook, New Jersey, U.S.A., on 11 April, 1894. He graduated from Princeton in 1916 and received his M.D. at Johns Hopkins in 1920.

During his undergraduate days Phelps became interested in human involuntary movement, as a result of some special work he undertook in physiology while at Princeton, and this laid the foundation for what was later to become his life's work.

His first internship was at Johns Hopkins, followed by another at the Massachusetts General Hospital, Boston, where he also did a term as assistant resident. In 1923, he became Orthopaedic Resident at the Children's Hospital, Boston, and then, in 1925, he was appointed to Yale as Assistant Professor of Orthopaedic Surgery, becoming Professor in 1929.

In 1936, Phelps left Yale to devote himself entirely to the problem of children with a motor handicap, and particularly those with cerebral palsy. He established the Children's Rehabilitation Institute in Baltimore, now well known throughout the world. Interested doctors and therapists visited the Institute from many countries; but the war of 1939–45 effectively hindered news of his work from entering Europe. However, on the cessation of hostilities, little time was lost, and the schools and clinics opened in the first few years (at least in the British Isles) all adopted Phelps's ideas and methods. As is almost invariably the case, these methods have become modified by other workers, but from my personal knowledge of Phelps I feel sure he would be the first to agree that this is on the whole a good thing.

Otherwise, Phelps is principally known to those concerned with the problem of cerebral palsy for his classification of the various infantile locomotor diseases hitherto largely regarded as separate entities, so that Little's disease, spastic diplegia, athetosis and choreo-athetosis came to be thought about under the generic term of 'Cerebral Palsy'. Many of us now disagree with his classification of the kinds of athetosis as being unnecessarily complicated and likely to confuse the issue; but, if Phelps is ever forgotten for his actual work, he will always be remembered as the man who made interested doctors, therapists and teachers think and think hard about a subject which had hitherto been incomprehensibly neglected.

The Children's Rehabilitation Institute and The Johns Hopkins Hospital have established an affiliation which will become effective when the Institute obtains a new building on a site adjoining the Hospital. The building is not yet started, but Dr. Russell A. Nelson tells us that both the Institute and the Hospital hope to see it in action in the next few years. This must surely be a matter for gratification to the founder and for congratulation from everyone working with handicapped children.

EDITORIALS

The American Pediatrician

Two very different views about the proper function and training of the American version of the pediatrician are put forward in the *American Journal of Diseases of Children* for November 1960. S. Z. LEVINE (p. 651) and CHARLES D. MAY (p. 661) both diagnose a sickness of the specialty itself, in a society which has largely eliminated the major ravages of infective and nutritional diseases. Both, in the true tradition of American medicine, make clear-cut recommendations for energetic treatment; and the fact that the treatments they recommend are diametrically opposed to each other follows logically from the differences between their diagnoses. Such differences need be no surprise when one considers the extraordinary variety exhibited by this, as by almost every other, aspect of the American scene at the present time.

Dr. LEVINE focuses his attention on the malaise, revealed by more than one survey, of many pediatricians in private practice. The great majority of the children whom they see 'are suffering from colds or stomach-aches, or are in good physical health': and their training has neither equipped them technically nor prepared them emotionally for dealing with such mundane problems. Dr. LEVINE accordingly prescribes a reorientation of the would-be practising pediatrician's training, away from the drama of the complex medical or surgical case, towards increased emphasis on emotional and social problems, towards contact with the patient in the home, and towards 'more day-to-day dealings with mentally retarded, physically handicapped and emotionally disturbed children, increasingly the real problems in today's pediatric practice'. He also mentions the need for 'more continuous experience with medical-related personnel—a veritable army that includes, among others, public health nurses and social workers'.

Dr. MAY, on the other hand, suggests that the cause of the trouble lies in the fact that the pediatrician tries to cover far too wide a field of activity. He prescribes 'a premeditated transfer of the bulk of ordinary care to better-trained general practitioners and more adequate public health agencies, while assuming greater responsibility for improving the training of these groups'. This would make it possible to concentrate on 'directing the efforts of fewer pediatricians to the roles of consultative specialists, teachers and investigators'. In other words, he favours a pattern of medical care of children more like that which has developed in Britain. But he takes the logical further step of recommending that the post-graduate training of the specialist pediatrician should be suited to his future function, being different in kind, and not merely in length, from the training in child health and diseases given to the future general practitioner. He also places significant emphasis on the role of the full-time investigator,

engaged in 'sophisticated fundamental research'.

This aspect of pediatrics has already been developed, in the U.S.A., on a scale which it is impossible to envisage in Britain. In many American children's hospitals and departments more members of the medical staff are engaged in full-time research, or have only very light and intermittent clinical responsibilities, than are directly and continuously concerned with patient care. The remarkable scientific results of this arrangement can be judged by referring to pp. 669–798 of the same journal, and studying the summaries of the 129 papers which formed the Scientific Program of the 1960 meeting of the American Pediatric Society. The visiting British pediatrician may be unable to see the clinical application of some of the work reported at such a meeting, and may even be quite unable to understand some of the more 'fundamental' papers; but he cannot fail to be impressed by the scientific validity and importance of much of it, and to notice how much of it depends on first-hand experience of the laboratory as well as the hospital ward. This is explained by the fact that the best of the specialists in training are expected to spend at least one year in fundamental research of some kind (indeed many medical schools now insist on the completion of a research project as a requirement for what we would call qualification). And vast sums of money are available to support such work, since they cannot be used for many purposes which might compete with the normal commercial basis of patient care.

But some American specialist paediatricians are also interested in the scientific study of clinical phenomena, in an intensive manner which is assisted by the development of subspecialties such as pediatric neurology or cardiology, or by specialisation within very limited age-groups, such as the newborn or adolescents. This whole theme is well illustrated by the presentation of the John Howland Medal and Award of the American Pediatric Society to Dr. BRONSON CROTHERS (reported on pp. 657–660).

Clearly, both the methods of treatment suggested by Dr. LEVINE and Dr. MAY carry some risk of toxic effects to the suspected patient, the specialty of pediatrics. The Editorial on p. 649 criticises both and adopts a *laissez-faire* attitude which we tend to associate with social development rather than with medical treatment in the U.S.A. The editor believes that the suspected patient is not so sick after all; and that his health can be left to look after itself by the normal processes of organic development in response to the need of patients, so long as there is a continuing supply of well-educated (as distinct from rigidly-trained) doctors imbued with the attitudes of Life-long Students. While we would all gladly echo some of these sentiments, as paediatricians we cannot but remember how much the development of our specialty owes to the planning made possible by the introduction of the National Health Service. And those responsible for future planning could learn much from our American colleagues, particularly in the spheres of research and medical education.

G. A. NELIGAN

Blindness in Little Children

"The child who has had favourable opportunities for learning before nursery school age is able to take his place in a regular nursery school programme with sighted children. There have been significant advantages to both blind and sighted children by having blind children in sighted nursery classes. It must be recognised that no nursery school programme can make up to the child for the lack of opportunities in the early years of life or overcome all the problems created by limited experience and inappropriate handling."

THIS quotation comes from a study of 300 blind pre-school children born in Chicago, who have been followed up for 5 years.* From this group 66 were chosen by random methods for intensive study. This included more than 2,500 psychological tests and more than 2,000 interviews between the workers and parents. Of the blind children of nursery school age over 75 per cent are actually doing well in sighted age-appropriate school environments. Of the children studied intensively 56 were blind from retrolental fibroplasia and 10 from other causes. The report shows the percentage of children at each age level passing the various items of the Cattell infant intelligence scale used. It also shows what proportion of children passed the social maturity tests. The development of blind and sighted children is compared, but with proper caution. Talking starts at about the same time in blind and sighted; socialisation is slightly late in the blind and they are later in holding a cup. Many of the blind children crept, although there has been an idea that blind children do not move about in this way. 'Walking assisted' and 'walking alone' tended to be slightly late in the blind. The overall functioning level of the blind and sighted children was much the same. It is important to notice, however, that in blind children retarded functioning was not necessarily evidence of a basic mental defect. The use of numerical scores, such as mental age and I.Q. and social age, were of limited value in understanding the blind children's capacity. From their experience the authors conclude that where a child is not functioning at his age level, repeated testing is desirable before drawing a conclusion as to his capacities.

Where blind children had other physical handicaps, their performance was significantly below that of children with uncomplicated blindness. The distributions of ratings for children who were blind from other causes were less favourable than for the children with retrolental fibroplasia. Only 10 children blind from other causes were studied in this project. Since blindness of prematurity is rare, one would like to hear about the progress of more children blind from other causes who have grown up in these helpful conditions.

The staff attempted to study the degree to which these children reached

* Blindness in Children. By M. Norris, P. J. Spaulding and F. H. Brodie. *Chicago:* University of Chicago Press, 1957, pp. 173, 22*s.* 6*d.*

optimal growth; they concluded that the basic characteristics of optimum growth were independence, initiative and active interest in the surroundings, all of which present difficulties for blind children. Four major headings were used in making a summary of impressions of the interview with the child: the family relationships, the child's opportunities for learning, the child's functioning, and the prognosis for the child. The *opportunities for learning proved a most important item as regards the prognosis* for the child's total capacities later on. The authors note that the so-called negativistic period, never an easy one for any child, is intensified for the blind child. What the children achieved was *not* very closely correlated to the degree of blindness. Some children with extreme retinal damage were still able to function visually. The use of vision was related to the general level of the child's functioning and not to what the ophthalmological examination revealed. Even the ability to get about did not appear to depend directly on the degree of vision. The totally blind child is particularly dependent on favourable opportunities for learning if he is to move about freely. The degree of vision may not be as important as the parents' feeling about it and the way their feeling affects their relationship with the child.

The authors draw some important conclusions. They considered it impossible to identify the conditions under which optimum development of the blind child takes place, so that he grows into an independent responsible freely-functioning child. Favourable opportunities for learning, as these authors understood them, are more important in determining the child's eventual functioning than his degree of blindness, his intelligence measured by psychological tests or his parents' social, economic, or educational background. There seem to be no special problems attributed directly to blindness. The idea that developmental problems are caused directly by blindness obscures an understanding of the nature and treatment of the child's problems. The *earliest* years of the blind child's life and the pre-school years are of primary importance. They determine the course of his later development. The damage done by prolonged failure to satisfy a child's needs in his early life is apt to be permanent. The blind child needs an individualised approach with many disciplines mobilised. Failure to provide the essentials to healthy personality development may result in grossly retarded functioning and extreme emotional problems which do not respond well to treatment.

The deeper knowledge we have gained in the last few years about the relationship of the mother and child in the earliest years of his life is beginning to affect our management of children. It is now uncommon for children under 5 to be sent away to Sunshine Homes. Two or three years ago it was thought necessary for children of 2, 3 or 4 years to be sent away because they came from such 'disturbed homes'. Surely the answer, so far as the future is concerned, is that we should prevent these homes, from which a blind child comes, from becoming disturbed. The disturbance is secondary to the child's blindness. This involves early recognition both of the blindness and of the problems that arise for the parents of the child who is blind. It is commonly difficult to say in the early months of life whether a child is or is not significantly blind. During this period of doubt it is tremendously necessary to give the parents support. It is for us to

enable the parents to give their blind children the very best opportunities and the full emotional support that all children need for optimal development. And ophthalmologists who are asked whether a child can see well must be able not only to test his sight but to give time to the parents' troubles and be aware of their special needs as well as those of their child, or else to enlist the co-operation of a willing paediatrician.

R. MAC KEITH

Blind children at play

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Sturge-Weber Syndrome

THE struggle to keep abreast with contemporary publications, even in fairly restricted fields, is indeed a hard one for many of us. It so often seems to be a process of 'one step forwards, two backwards'—a battle that is never lost but where victory is never in sight. It is not surprising, therefore, that much of what is published commonly meets with a certain cynicism among its readers, who may adopt a highly critical attitude and quickly resent the seeming waste of time and space caused by 'unhelpful' papers. How pleasant it is, therefore, to see a monograph* which it is almost impossible to fault, which reveals the undoubted application, interest and wide knowledge of its authors, and which ends with definite and reasoned guidance on surgical treatment.

The Sturge-Weber syndrome is relatively rare, but its clinical features of epilepsy, mental retardation and (usually) hemiparesis are familiar to those caring for brain-damaged children. As ALEXANDER and NORMAN describe in their monograph, the increase of mental retardation following a series of epileptic attacks is notable in this disease and usually leads to the patient's admission to an institution, where he may die at an early age in status epilepticus.

The authors have considered the many published reports, but most of their information has been derived from the very careful study of 7 cases of their own, 5 of which were operated on. Excision of the angiomatose cortex, quite superficially, was apparently not an unduly difficult or hazardous procedure in ALEXANDER's hands and was followed by very definite arrest of the patient's deterioration and in time by a remarkable improvement in mental ability. The seizures are reduced or abolished by operation.

The deterioration in mental ability which occurs sooner or later in sufferers from Sturge-Weber disease is closely related to the incidence of fits, sudden bursts of which may leave the patient remarkably impoverished. This phenomenon has impressed ALEXANDER and NORMAN who nevertheless realise that other processes probably play a part in the downhill course of the disease. They have been convinced, however, from studying their own cases that excision of the angiomatose cortex is not only helpful but almost obligatory if the patient is to be helped at all. The increasing inefficacy of drugs as time passes encourages one to agree with this opinion. It is difficult to know how early in life to operate (ALEXANDER and NORMAN suggest not later than 2 months of age), and the decision to do so will demand courage in both parents and medical advisors.

At the very least, this monograph will stimulate further interest in Sturge-Weber disease, and its constructive conclusions may well point the way to much alleviation of suffering.

KENNETH TILL

* The Sturge-Weber Syndrome. By G. L. Alexander and R. N. Norman. Bristol: John Wright, 1960, pp. 95, 32*s.* 6*d.*

Breast-feeding in New Zealand

THE surveys of breast-feeding in New Zealand published by the Plunket Society in 1938–39, 1944–45 and 1951–52 showed a steady decline in both the establishment and the duration of breast-feeding. The Plunket Society and the University of Otago have since made a joint study[1] of nearly 900 women confined at one hospital in 1952–54. During this period the antenatal and postnatal supervision of infant feeding was under the direction of a nursery sister trained in the techniques of the late Harold Waller. Under this regime lactation was established in a higher proportion of mothers than the national average. However, a follow-up did not show that this higher level was maintained. This was attributed mainly to unexplained milk failure and secondly to the fact that 15 per cent of mothers left hospital giving complementary feeds.

In a broader survey of breast-feeding —broader in both time and place— Dr. McGeorge[2] first considers the changing incidence of breast-feeding in all parts of the world. The rate of decline is roughly proportional to national prosperity, but the incidence remains high in Scandinavia, is influenced by the premium payable in Switzerland, and is far lower in the westernised than in the poorer parts of India. A brief historical survey of infant feeding reminds us that a substantial proportion of mothers have always been reluctant to breast-feed when alternatives were available. The reasons underlying the widespread decline in breast-feeding are discussed in terms of physical and social impediments. Dispassionate examination of the relative values of breast- and bottle-feeding fail to reveal any convincing evidence that either breast milk or breast-feeding have any great advantages over other foods and methods. Dr. McGeorge reasons that time is well spent in the antenatal clinic and lying-in ward ensuring that every mother has the best opportunity to breast-feed; but when this proves impossible or unsatisfactory there should be no delay in having recourse to other methods of nourishing the baby.

A high incidence of breast-feeding is encountered only when there is no available alternative, or when mothers have been led to believe that breast-feeding carries some nutritional or psychological advantage to the baby. For centuries mothers have been exhorted to breast-feed their babies, implying that they have always been reluctant to do so. The present-day failure to find any advantage accruing to the breast-fed baby may simply reflect a failure to look in the right direction. Imposing though the literature on infant feeding is, the last chapter has not yet been written. Dr. McGeorge's paper is an up-to-date, well-informed and comprehensive survey of a fundamental paediatric problem and records without emotional bias the state of affairs in 1960.

R. W. SMITHELLS

1. Deem, H. and McGeorge, Murray. 'Breast-feeding.' *N.Z. med. J.*, 1958, 57, 539.

2. McGeorge, M. 'Current trends in breast-feeding.' *Ibid.*, 1960, 59, 31.

Amino-acid Excretion in the Urine

SINCE the clinical application of partition chromatography as a method of separating amino-acids by DENT in 1946, increasing interest has been taken in patterns of excretion in the urine. It has become evident that some specific patterns are associated with recognisable conditions, such as phenylpyruvic oligrophrenia, H-disease, and alcaptonuria.

Amino-acids are needed in the body for various purposes besides conversion into tissue protein; those that are absolutely necessary for this function being called 'essential' amino-acids. Some are precursors or constituents of vitamins, coenzymes, or hormones. For example, phenylalanine may be converted into tyrosine and then either passed into the channel of protein formation or used in the synthesis of thyroxine and epinephrine, or in the formation of melanin. Tryptophan, an essential amino-acid, is a direct precursor of nicotinamide and serotonin. Some amino-acids are important in carbohydrate metabolism, being necessary for the formation of enzyme systems in the intermediate stages. Indeed, it has been said that most if not all congenital metabolic disease may be attributed to abnormalities of protein synthesis.

The source of amino-acids is the enzymatic breakdown of dietary protein in the gastro-intestinal tract. There appears to be active absorption by the mucosal cells. Thence transport is by way of the portal system to a state of equilibrium with the various tissues. The blood-level is fairly stable, though there is a slight rise after a protein meal.

Amino-acids entering the renal tubule by filtration through the glomerulus are mostly reabsorbed by the tubular epithelium, probably by various mechanisms. This is indicated by the fact that some are reabsorbed in competition with others and some independently. It is likely that essential amino-acids are absorbed preferentially. Other substances, such as sugars, may also compete. Thus in galactosaemia there is an aminoaciduria which is cured by the administration of a lactose-free diet, though this in no way influences the basic enzyme defect.

Diseases with organic aciduria may be classified as follows:

1. Blood-level of amino-acids raised, with no evidence of renal abnormality —e.g., liver disease, prematurity, phenylketonuria, and wasting diseases such as extensive muscular atrophy. Neuronal disease is not in this category.

2. Blood-level of amino-acids normal; failure of reabsorption in states where there is renal disease (including those caused by toxic agents, such as excessive dosage of vitamin D), or pituitary or hypothalamic disease with polyuria.

3. Blood-level normal, with no demonstrable renal disease.

The third group is probably the most interesting and important clinically. In the first two mechanisms the aminoaciduria is secondary to the existing disease, which is probably evident clinically. The third mechanism—primary aminoaciduria—requires full chemical investigation to provide enough information for a differential diagnosis.

An association between congenital defect, mental retardation and aminoaciduria is becoming recognised, though of course not everyone with aminoaciduria is mentally retarded. There is the suggestion, however, in these various conditions, of a common basic metabolic abnormality. Not many years ago it was sufficient for a doctor to diagnose mental retardation and dismiss the subject, since no treatment was possible, a situation comparable to congenital heart disease before modern surgical and anaesthetic techniques became available. The tool of paper chromatography opens up wide areas in what was previously an unpromising and discouraging field of clinical material.

Before disease states can be recognised, however, the normal patterns must be known. Excretion varies with age. As long ago as 1911 it was recognised that the amounts of alpha-amino nitrogen (representing amino-acids) excreted in the urine were much greater in infancy than in adult life. There is a rapid change in the pattern in the first week of life, followed by a more gradual change over the next five months. This probably reflects the initial renal immaturity. In addition, it has been found that there is a wide range of excretion in apparently normal people within the same age-groups, though siblings tend to have somewhat similar patterns of excretion. To obtain standards of normality one must examine very many samples of urine. This is not such an attractive task as the description of some new syndrome. Those who do this tedious work deserve our encouragement and thanks.

When the normal is known, the finding of abnormal aminoaciduria will be the first step in unravelling the steps of a metabolic process with the hope of treatment. This might take many forms —perhaps dietary restriction, as in phenylketonuria; or such procedures as the introduction of cultures of organisms to replace the missing metabolic process, such as the conversion in the bowel of phenylalanine to tyrosine. Such possibilities are exciting.

J. N. MONTGOMERY

ORIGINAL ARTICLES

The Care of Children with Motor Handicaps in Sweden

MARCEL D'AVIGNON

Paediatrician to Eugeniahemmet and the Central Swedish Hospital and School for Children with Motor Handicaps, Stockholm

THE care of the disabled in general, and of children and young people with motor handicaps in particular, is a long-standing tradition in Sweden. The oldest of the five institutions for the disabled, Eugeniahemmet, was founded in 1879 by Princess Eugenie. The others are the Homes for the Disabled in Hälsingborg, Gothenburg, Stockholm and Härnösand. Eugeniahemmet admits children only, and these come from all over Sweden. The Homes for the Disabled, on the other hand, take patients of all ages from local areas.

Paediatric Care

The care of sick children in Sweden is based on about 40 paediatric departments, each well organised under a highly qualified paediatrician often with professorial status. These paediatric units include the six university clinics, at Stockholm (2), Uppsala, Lund, Gothenburg and Umeå.

Care of Motor-Handicapped Children

Congenital diseases or malformations resulting in motor handicaps are not notifiable in Sweden, nor are diseases or accidents which later give rise to such handicaps. A child with a motor handicap will in the first place be admitted to the paediatric unit nearest his home-town. The great majority of confinements take place in hospital so the child will be cared for initially in the paediatric department of the hospital where he is born. The disabled infant may be transferred subsequently to the orthopaedic department of a general hospital, to the orthopaedic university clinics or to the Homes for the Disabled which have orthopaedic departments with surgical facilities.

The children are otherwise cared for at home. Only more chronic cases, which are too difficult to handle or in which social conditions make home care impossible, may stay for longer periods in the paediatric departments or, in a few cases, at Eugeniahemmet.

Some of the children's clinics in Sweden have also so-called B-departments for the care of the chronically sick child. There are no other special homes for chronic child cases. If after observation in a paediatric department a child is found to be feeble-minded, he is admitted to an institution for the feeble-minded if the parents are for one reason or another unable to take care of him. A few children with motor handicaps may be placed in Children's Homes, but these are not intended for such cases, and it is only pending another placing that a handicapped child may be temporarily admitted to a Children's Home. There is thus no special organisation for pre-school children with motor handicaps, except for certain well-defined groups, such as the blind, the deaf and the cerebral palsied, and in a certain number of cases those who have had poliomyelitis. A child with, for example, *spina bifida*, with paraplegia, progressive muscular atrophy, arthrogryposis etc., is

thus not covered by any special organisation other than that of the hospitals and general social care.

Education

Compulsory school attendance begins in Sweden when the child is 7 years of age. If parents have a motor-handicapped child there is nothing to prevent their keeping him at home. They are entitled to home tuition up to one hour per day at the expense of the local authority. Since, however, the latter have no special funds for this, an attempt is generally made to send the child to some established form of school. If he is mentally under-developed, a year's postponement is usually allowed. If the child cannot keep up in a normal school, he is placed in a special school administered by the official authority for the care of the feeble-minded, or in a Home for the Feeble-minded. There are special schools for blind and deaf children as well as those suffering from cerebral palsy.

If, however, the child is normally gifted or just below average, so that he can benefit from tuition in the ordinary elementary school, with its normal and supplementary classes, this must be arranged accordingly. If the child's motor handicap is such that he can be tended by a local physician or at the nearest children's or orthopaedic department, while living with his parents, his schooling is, if possible, arranged in the home-town.

If, on account of distance or for social reasons, this is not possible, the child may be placed in one of three schools, at the Homes for the Disabled in Hälsingborg, Härnösand or Eugeniahemmet in Stockholm. The last-mentioned is the biggest, with 150–160 places. Here the children can also obtain the necessary medical attention. If it is found that such children cannot benefit from this tuition, transfer to schools under the administration of the authority for the care of the feeble-minded is still possible at this stage. The schools at the Homes for the Disabled in Hälsingborg and Härnösand each have places for only 30 children, and the less severe cases are admitted here, the severer cases, on the other hand, being taken by Eugeniahemmet.

The schools are run on the boarding school basis, but children living nearby may attend as day-pupils. In these schools the syllabus conforms as closely as possible to that of the normal school, though standards are of course not so exacting, and teaching is necessarily a good deal more time-consuming and individual. Schooling is completed at 15 years of age as a rule but the stage of maturity rather than the biological age is taken as the criterion for this. If the child is immature a further year at school may be suggested. For many children at this juncture a pension is necessary, and the qualifying age for this is 16 years.

During the last year at school, as in all schools in Sweden, vocational guidance is given, vocational tests are carried out, and in some cases the child later receives a course of training for some occupation. Vocational tests are held under the aegis of the State authority for rehabilitation, which has branches in different parts of the country; in the case of Eugeniahemmet the testing is done at the National Rehabilitation Clinic. The following possibilities are open. Children who show promise in their studies may continue these up to the lower-certificate examination at the Institute for the Disabled in Stockholm. Those that can go further to the matriculation examination and higher studies go to high school and university. They may then stay on at the Institute for the Disabled, but as a rule they are boarded in a home in which the necessary special care is available. The other category of young people whose motor handi-

cap and mental development allow them an occupational training, can get this in the occupational schools of the Institutes for the Disabled. If, at any Institute for the Disabled suitable training facilities are not available, the pupil can attend the local training school for normal children. In this case the child may either stay on at the Institute for the Disabled or be put out to board in some suitable home.

The third, more severely handicapped category may learn some suitable hobby in a general workshop, more as an occupation and rarely to provide more than a partial livelihood. As a rule these children are kept at home as long as this is possible for the parents; otherwise, like the fourth category of the most severely handicapped, they are placed in a special nursing home. These homes for the chronic sick are administered by the county councils and admit all forms of chronic disease irrespective of age. The policy is to place the young people near to the town of domicile, so that relatives can visit them as often as possible. What we now need in Sweden are nursing homes specifically for younger chronic cases, and their organisation is at present under consideration. Smaller homes of this kind for the motor handicapped would be desirable, provided young people are not placed too far away from their place of domicile.

All schools and the various hostels are free or charge very small fees. Parents of children cared for at home are eligible for state aid in respect of medical care, rent and, up to the age of 16 years, the ordinary child allowance. After the age of 16 years the patients living in their homes may themselves get a pension.

Community Aid

The modern Swedish community tries to plan to help the motor handicapped. There are lifts for invalids' chairs, banisters and escalators in many public buildings. In new residential areas suitable flats are built for the handicapped; kitchen furnishing, water closets and bathrooms are planned individually, so that the handicapped housewife can manage the household by herself.

Eugeniahemmet

The care of children and young people with cerebral palsy has been organised as described; but Eugeniahemmet has been particularly concerned with the care of cerebral palsied children because of the interest orthopaedic physicians employed there have taken in these cases. Since the beginning of the 1950's the care of cerebral palsy has begun to strike out on more individual lines in Sweden, coincidentally with the increased interest evident both in America and in Europe. Here, too, the importance of early diagnosis was realised, as also the value of early treatment in place of the older practice of postponing this until only orthopaedic operations could be attempted with a view to improving the condition.

Especially in Stockholm, Uppsala and Gothenburg the paediatric university clinics began to show increasing interest in cerebral palsy. Cases were diagnosed earlier, and children then admitted with their mothers if they lived at a distance; otherwise they were treated as outpatients. The training of students, doctors and nurses was intensified, particularly with a view to early diagnosis. Gradually interest has spread, and an increasing number of children's departments provide special staff to treat these children. Smaller kindergartens and homes, both of the boarding and day-school type, have been established in many parts of the country.

In the meantime, *Eugeniahemmet* has been re-organised to become the central institution for handicapped children from all over Sweden. To the original staff (which included an orthopaedic surgeon),

a children's physician, a children's psychiatrist and a phoniatric physician have been added. An ear, nose and throat specialist, an audiologist, ophthalmologist, EEG-specialist, and if necessary a neurologist and neuro-surgeon are now available. X-ray and laboratory investigations can be performed. The number of physiotherapists has been increased and a special teacher for the deaf engaged, both for the small children and for the school. Special speech therapists have been employed and likewise special kindergarten teachers and social workers. At the same time an older department in the home has been modernised to provide 40 beds for children aged 1 year and upwards. In other words, the home has been equipped for completely up-to-date cerebral palsy care.

Nevertheless, it remains a desideratum for small children to be cared for as near as possible to the parental home, however warm and home-like the hospital milieu may be. For this purpose small cerebral palsy centres have been opened in most large towns. These centres are very well equipped and all have access to consultant paediatricians, children's psychiatrists, orthopaedic surgeons, etc. Thus Gothenburg, Malmö, Uppsala and many other towns have day-schools, while Orebro and others have boarding-schools, and many centres have pre-schools.

Where it has not been possible to give the treatment at or near the child's home and it has been necessary to admit the child to Eugeniahemmet, this has been done in periods of a couple of months at a time, as a rule twice a year. Then when the child is discharged the mother is given detailed instructions from all the staff who have treated the case; she receives necessary aids such as boots, inserts, bandages, invalid chairs, go-carts, etc.

Orthopaedic operations are also performed, though on a much smaller scale than formerly. There are case discussions between all the staff where necessary, and with the help of the psychologist, the psychiatrist assesses the mental development and discusses the problems with the parents.

The small children in the institution receive different forms of therapy daily. The older children at the school have less time for physiotherapy, their day being already taken up with school work, free-time occupations, perhaps special hearing lessons or speech therapy. They are usually treated every other day. In the paediatric department the clientele consists, with a few exceptions, of cerebral palsied children; but at the school there are approximately 60 per cent of cerebral palsied children, the remainder being cases of muscular dystrophy, old poliomyelitis, *spina bifida* with paraplegia, chronic rheumatoid arthritis, arthrogryposes, skeletal malformations, some chondrodystrophies etc.

There is no special form of occupational training for the different categories of the motor handicapped. The cerebral palsy cases represent a smaller percentage of those who learn a trade or who continue with their studies. This is of course due to the fact that their motor injury is often severe and that psychic changes are likely to preclude studies beyond the elementary school stage. Children suffering from cerebral palsy are often slow and have difficulty in concentrating. This, combined with their motor difficulties, makes occupational training hard. Like all other motor handicapped cases, however, they are nevertheless given a trial within the frame of state rehabilitation.

There are two categories of cerebral palsied children who are not admitted to Eugeniahemmet, but who remain at home. Either the damage is so slight that they are able to attend the ordinary school in their home-town or one of the new cerebral palsy centres, in which case they will

probably go on to a higher school education and occupational training in some form and be able to earn a living on their own. Or they belong to the category of severely injured children whom the parents are unwilling to relinquish; these will probably be placed directly in institutes for the feeble-minded or in nursing homes for chronic cases as soon as it becomes impossible for the parents to look after them at home.

Future Outlook

The care of cases of cerebral palsy is at present being reorganised. In a large number of children's departments in our hospitals investigation and care as well as special treatment is carried out in accordance with the principles described above. In these departments there are also kindergartens where the children have play therapy every day, as well as physiotherapy. In several parts of the country there are also schools in which children suffering from cerebral palsy receive ordinary tuition at the same time as treatment of different kinds. In Gothenburg, Bräcke Östergård is a newly built, extremely pleasant and well designed home, where 30 children with cerebral palsy are admitted from western Sweden from the ages of 1 to 7 years. A boarding-school for 30 school-children, corresponding to that at Eugeniahemmet, although it will be more up-to-date and designed specifically for cerebral palsied children, is ready now. It has also recently been decided that a new cerebral palsy school both for boarders and day pupils shall be built in Uppsala: the Folke Bernadotte School. The premises have been well planned on the basis of experience of other institutions for handicapped children. Stockholm is too remote for the major part of Sweden, and it is often undesirable to send children too far from home; smaller boarding schools in different parts of the country would therefore make things easier for parents. The regional cerebral palsy schools discussed above would chiefly admit less complicated cases. According to the new organisation Eugeniahemmet would then take care of cerebral palsied children from the largest district in the country, the Stockholm area, as well as those with severer cerebral palsy combined with special defects, of speech and hearing etc., from all over the country.

Interest in cerebral palsy is at present focused on the young. Very few of these, or at all events of the severest cases, can compete effectively in the community. For those who have an occupation, sheltered workshops are needed. Such workshops do exist in Sweden in the bigger cities, but the cerebral palsy clientele in them is small. For the most severely handicapped cases Stockholm has a workshop which is state-aided and not self-supporting. There is also a department for chronically ill children and young people, including a number of cases of cerebral palsy, which differs from a nursing home inasmuch as old people are not admitted. A hostel for chronic cases in young people is being planned in Stockholm as an annexe to a youth hostel. This department has its own workshop for severely handicapped adolescents. This is the first project of its kind in Sweden and will doubtless be followed in the larger cities.

SUMMARY

The medical and educational care of motor-handicapped children and young people in Sweden is described. Emphasis is on home care where possible. Regional and local institutions offer specialist treatment and educational facilities, and provide residential accommodation if desirable.

Eugeniahemmet in Stockholm is a national institution of 160 beds which provides modern comprehensive care of the cerebral-palsied child.

Re-organisation of the provisions for cerebral palsied children is in progress, *pari-passu* with an increased national awareness of the problems of the handicapped.

RÉSUMÉ

Soins aux enfants atteints d'infirmités motrices, en Suède

Description des soins médicaux et éducatifs donnés en Suède aux enfants et aux jeunes gens atteints d'infirmités motrices. La préférence va aux soins à domicile, partout où ils sont possibles. On trouve dans des établissements régionaux et locaux des traitements par spécialistes et des facilités d'ordre éducatif; on peut y séjourner, si c'est opportun.

A Stockholm, Eugeniahemmet est un établissement d'Etat de 160 lits où l'on donne des soins modernes éclairés à l'enfant infirme moteur cérébral.

La refonte des dispositions relatives aux enfants infirmes moteurs est en cours, cependant que la conscience nationale s'émeut de plus en plus des problèmes de l'infirme.

ZUSAMMENFASSUNG

Die Pflege der Kinder mit motorischen Störungen in Schweden

Ärztliche und erzieherische Pflege der Kinder und Jugendlichen mit motorischen Störungen in Schweden wird beschrieben. Wo möglich ist, hat Hauspflege den Vorzug. Regionale und lokale Einrichtungen bieten Behandlung durch Spezialisten und Erziehungserleichterungen und beschaffen Wohnmöglichkeiten, wenn es wünschenswert ist.

Eugeniahemmet in Stockholm ist eine Staatsanstalt von 160 Betten, wo das Kind mit Zerebrallähmung sachkundige moderne Pflege bekommt.

Die Reorganisation der Einrichtungen für Kinder mit Zerebrallähmung ist im Gang, gleichzeitig mit dem zunehmenden nationalen Bewustsein der Probleme des Krüppels.

Phoniatric Treatment of Children with Cerebral Palsy

ELLY OHLSSON-EDLUND, M.D.

From the Eugenia Home, Stockholm (Med. Superintendent: H. Nilsonne, M.D.)

The Eugenia Home (Eugeniahemmet) in Stockholm is a boarding-school for children with motor handicaps. In addition, there are two sick wards with 40 beds. The staff includes an orthopaedic surgeon, a paediatrician, a child psychiatrist, a medically qualified speech-therapist, a dentist, and nurses and medical gymnasts.

THE school covers the Swedish elementary system, with both normal and auxiliary classes. There is also a kindergarten in the medical division. The Home, however, does not have facilities for intellectually retarded children. There is a special division for the training of hearing, as well as a reading clinic for children with special problems in reading and writing (dyslexia and dysgraphia). Children with speech disorders receive 'phoniatric' treatment, carried out by a speech therapist, together with an elementary school teacher and a specially trained nursery school teacher. At the time of their general medical examinations, children with speech difficulties are referred for specialised phoniatric examination. The school teachers give their opinion on the need for speech therapy and on this matter the child's parents may also be consulted.

Examination

The method of examination and treatment varies widely, according to the child's age and the type of speech defect. In small children it is useful to make a short first contact in the presence of a nurse whom the child knows well. It is important for the children to have confidence in the therapist and they must understand and feel secure in their new surroundings before they can respond to therapy. They sit during the examination but may either sit or lie down during therapy.

At the examination, the therapist notes the degree of speech development and understanding, reaction times, voice pitch, spontaneous speech compared with attempts to pronounce sounds and words, speech rhythm, motor difficulties of the lips, tongue, jaws, floor of the mouth, gums, pharynx, trachea, the carriage of the head, breathing, accompanying body movements, etc. A general impression of sight, hearing and psychological development is recorded, but specialists in these fields are also consulted.

Therapy

The *special phoniatric equipment* includes a series of tuning forks from CI to CV. These help to give an idea of a child's hearing, but their phoniatric use is to make a listening child pay attention. CI is struck and the fork placed against a table, a little distance away from the child. Often the child will then look up, listen and smile. Soon the tuning-fork can be put to the child's ear. In this way C, CI and CII are tried and followed carefully by CIII, CIV and CV. Care is necessary because more than half the children reported below preferred CI to CV, which some

children say they don't like. A few children prefer Cv, while others have no definite opinion.

The children are encouraged to make vocal sounds at different pitches of the tuning-fork. They are assisted in finding a suitable vowel, for example a phonetic U or O at low pitches (CI or CII) and then, with or without help, they find phonetics suitable for Cv. Often the child can hold the vowel sound for only a short time. The attempt, however, is continued as a game, and the child works up his phonation time during a course of treatment from, say, a quarter of a second to five to ten seconds. He is encouraged to make as free vowel sounds as possible, thus improving the sound of his speech. When the child has got used to the tuning-forks, a ringing fork (C—CII) may be placed against the crown of his head, while his hearing is cut off by stopping his ears. Often the child then 'listens' intently and with pleasure.

Tuning-forks from CI (32 vibrations) to CII are used further for *vibration therapy*. The CI fork is struck and the child is soon capitivated by watching it vibrating or standing still. Soon he makes an attempt to start the fork himself. In this way children are prepared to accept vibration therapy for tension of the speech musculature. The attempt is continued by placing one limb of the vibrating fork against the child's upper lip and encouraging him to say ba or be. Thus the muscle structure is activated, and as the child feels the vibrations he makes the desired labial sound more correctly or more easily than before.

Later, the tip of the child's tongue can be activated by vibrations transferred from the tuning-fork via a narrow wooden stick, such as an ear probe. The stick is placed with one of its ends against the underside of the tip of the tongue and the other against the outside of one limb of the fork. This may be used for practising the rolled 'r'. Vibration therapy can then be gradually extended to other speech muscles.

Other forms of therapy consist of gentle *manual movements* which aim at forming the child's speech organs for the normal movements and helping to produce relaxed speech and a natural carriage of the head. The supine position is used to facilitate relaxation while breathing is largely practised lying down. Spatulas are used for practising movements of the tongue. The child is also encouraged to develop active tongue movements by trying to 'push' away the wooden stick which he feels against his tongue. Gentle pressure with the thumb under the chin, with the child's lips lightly closed, is sometimes used, especially for the l-sound. This method is applicable to cases in which the tip of the tongue tends to point outwards instead of upwards which occurs when the tonus is greatly increased. Various little tricks are used such as getting the children to blow feathers in practising hissing sounds, which may be difficult for as many as 70 per cent of children.

Finally, we used certain *materials* for attracting the child's eye, including light wooden blocks about $3\frac{1}{2}$ inches square and $\frac{1}{4}$ inch thick. On one side, the block has a picture in clear bright colours, of the mouth forming a certain sound, such as *u*, *o* or *s*. On the other side there is a picture of an animal or fruit, whose name begins with the same sound, together with the corresponding initial letter. The children can sit looking at the blocks, turning them over and comparing them, either on their own or with a little help, at the same time feeling that they have learnt something for themselves. Mirrors are also used in suitable cases for training in articulation and for refining speech movements. All speech therapy is given with the greatest possible encouragement. The

child should feel that he himself has done something right or almost right. For the same reason the children are allowed to hear the parts of tape recordings on which they themselves can appreciate their progress.

Results

The following results are based on 50 children (31 boys, 19 girls) with cerebral palsy treated according to these principles, and all patients in the Eugenia Home who received speech therapy between 1958 and 1960. They ranged in age from 3 to 18 years and were distributed with regard to school classes as seen in Table I.

Cases of severe deafness are not included, but 11 cases of perceptive or nerve type were noted. Ability to understand language largely corresponded to age, intellect, general development, and experience. The main diagnosis was spasticity or athetosis in about equal numbers, and there were a few cases of ataxia. There were also combinations of spasticity and athetosis, and of ataxia and athetosis. In many cases shifts of tonus were noticeable, while impairment of movements was often severe.

The different types of speech defect treated are seen in Table II:

The results of treatment of these groups is shown in Table III:

Of the 32 school children, 24 had also received periodic speech therapy during their earlier period at the Eugenia Home and thus may have received speech therapy during the whole of their stay in school; 14 of these are included in the group showing considerable improvement and in

TABLE I
NUMBERS OF SCHOOL CHILDREN AND PRE-SCHOOL CHILDREN IN DIFFERENT CLASSES

School children (32)	Normal class	15
	Auxiliary class	13
	Observation cases	4
Pre-school children (18)	Should be able to go to school	8
	Observation cases	10

TABLE II
NUMBERS OF CHILDREN WITH DIFFERENT TYPES OF SPEECH DEFECT

Group		*Nos.*
I	Retarded speech	32
II	Articulation disorder of cerebral palsy type ..	42
III	Voice disorders of cerebral palsy type	42
IV	Nasalising	14

TABLE III
ASSESSMENT OF SPEECH AFTER TREATMENT BETWEEN 1958 and 1960

Group	*Considerable Improvement*	*Improved*	*Slight Improvement*	*Not Improved*
I	9	15	5	3
II	4	15	19	4
III	12	13	11	6
IV	5	5	2	2

some cases almost perfect speech. The results bear no definite relation to whether the children were in normal or auxiliary classes. Each group is distributed about evenly between the two types of school classes.

One case deserves special mention. A girl, now 16-years old and almost completely disabled through athetosis and considerable variations in tonus, had, at the age of 10 years, a toneless, rigid voice and a phonation time of about $\frac{1}{4}$ second. At 12 she had a freer and clearer voice pitch, and now at 16 she has a beautiful voice with a phonation time of 4–5 seconds.

The *pre-school children* received speech therapy adapted to their ages over periods ranging from a month's observation to repeated treatment lasting 2 years. Their distribution is summarised in Table IV.

This comparatively small series does not permit any definite conclusions, but the results support the view that speech therapy should have an important place in the care of children with cerebral palsy and should be continued for long periods. Apart from the valuable contacts acquired in this way, the child profits greatly from the experience of having his speech trained and obtaining some result.

TABLE IV
RESULTS OF TREATMENT. PRE-SCHOOL CHILDREN

Duration of treatment	*Considerable Improvement*	*Improved*	*Slight Improvement*	*Not Improved*
2 years or more	2	2	—	—
1 year or more	2	2	—	—
About 5 months	1	1	—	—
About 4 months	—	1	—	—
About 3 months	—	—	1	—
About 2 months	—	2	—	—
About 1 month	—	1	1	1

SUMMARY

The phoniatric treatment of cerebral palsied children with serious motor handicaps and relatively normal intellects is described. Using tones from a tuning-fork series C—Cv the children notice the qualities of sounds. The speech muscles are stimulated with the help of vibrations from the tuning-fork, Ci to Cii. The children may look at pictures which show how certain sounds are formed and compare the mouth positions of the various sounds. They check in a mirror how they themselves articulate. Furthermore, they may hear their own speech on a tape recording. After long training the children in many cases really speak better.

RÉSUMÉ

Traitement phoniatrique des enfants atteints d'infirmité motrice cérébrale

Cet article est consacré au traitement phoniatrique des enfants infirmes moteurs cérébraux, atteints de graves infirmités physiques mais ayant conservé une intelligence relative. Les enfants notent la qualité des sons d'après la tonalité d'un diapason de la série C—Cv. Les muscles de la parole sont stimulés par les vibrations du diapason en Ci—Cii. Les enfants ont la possibilité de regarder sur des images le processus de formation de certains sons et de comparer les positions de la bouche à chaque son. Les enfants

contrôlent dans une glace leur propre articulation. D'autre part, ils peuvent s'entendre parler sur des bandes enregistrées. Les résultats prouvent qu'une longue éducation permet a de nombreux enfants de bien mieux parler.

ZUSAMMENFASSUNG

Phoniatrische Behandlung der Kinder mit Zerebrallaehmung

Dieser Artikel befasst sich mit der phoniatrischen Behandlung der Kinder mit Zerebrallähmung, mit schweren körperlichen Gebrechen, aber mit einigermassen erhaltener Intelligenz. Mit Hilfe der Töne einer Stimmgabel der Serie C—Cv, bemerken die Kinder die Lautwerte. Die Sprechmuskeln werden durch die Schwingungen der Stimmgabel CI—CII angetrieben. Die Kinder können auf Bilder blicken die zeigen, wie gewisse Laute gebildet werden, und die Haltungen des Mundes bei den verschiedenen Lauten vergleichen. Die Kinder kontrollieren in einem Spiegel wie sie selbst aussprechen. Ferner können sie ihre eigene Rede auf einem Band hören. Die Resultate zeigen, dass die Kinder nach langer Ubung in zahlreichen Fällen viel besser sprechen.

Specific Conditioning Treatment of Enuresis Nocturna*

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The Development of Bladder Control

AN adequate analysis of the aetiology and treatment of bed-wetting needs to take into account the development of voluntary and nocturnal control of the bladder. The automatic reflex micturition of the infant is in many ways similar to that of adults in whom disease or surgery has isolated the bladder from the control of higher nervous centres. Neural maturation provides connections between the cortex and lower centres which allow the development of the ability to voluntarily initiate or inhibit micturition, and conditioned reflexes are established which link this initiation and inhibition with various environmental situations. Thus, the older child chooses, within limits, the time and place at which he urinates and tends to experience the urge to urinate when in a 'toilet' environment or when adopting certain postures.

Nocturnal continence is very rarely achieved until bladder-control in the waking state is well established. This strongly suggests that similar mechanisms are involved but that the maintenance of cortical inhibition is more difficult during sleep than in the waking state. The sleeping state essentially involves the inactivation of the higher parts of the brain, especially the cerebral cortex, and, during sleep, the threshold for most somatic reflexes is raised but that for most autonomic reflexes is lowered. Certain areas of the cortex which control vegetative functions are described as 'sentinel points' and remain active during sleep. Other centres may become locally activated in response to appropriate stimuli without disturbing the general state of sleep of the organism (Delafresnaye, 1954; Hodge and Hutchings, 1952). Such 'sentinel points' for the inhibitory control of bladder reflexes are established as a result of neural maturation and day-time learning. With further maturation and increased potency of these centres, they are likely to remain locally active during sleep or to 'awaken' in response to bladder and postural stimuli.

The operation of these mechanisms is portrayed schematically in Fig. 1. The unbroken lines represent the course of events at the stage when the child is continent during the day but incontinent at night. Throughout the range of bladder pressures up to and even at the threshold for micturition, the cortical sentinel point for the inhibition of micturition is relatively inactive. Sleep may be somewhat disturbed, but the child urinates without waking and sleeps on in a soiled state.

When cortical influences have become more potent the sentinel point becomes, as shown by the broken lines, fully active well before the micturition threshold is reached. Micturition is inhibited and, as pressure increases, sleep becomes progressively shallower until the child wakes, experiences the urge to urinate, and goes

* This paper is a condensed version of part of a chapter to be published in *Behaviour Therapy and the Neuroses*. London: Pergamon Press, 1961 (Ed. H. J. Eysenck).

to the toilet. An increasing bladder capacity would be expected to result from anatomical growth and from the effects of the cortical inhibitory influences on the detrusor muscle tone. Ultimately, tolerance will be sufficient to enable the child to sleep throughout the night in normal conditions.

Aetiology of Nocturnal Enuresis

The literature concerning the aetiology of enuresis is extensive and theories numerous, but, without gross injustice to the authors' points of view: most theories may be subsumed under one or other of six categories:

(1) *Specific Physiological Dysfunction*

Some authorities, such as Crosby (1950), consider enuresis, not as a symptom of some more general disorder, but as a specific clinical entity. He uses the term 'essential' enuresis to describe cases of this type.

(2) *General immaturity*

Others, such as Kanner (1947), treat enuresis as one aspect of general immaturity, which may also be reflected in retarded speech development, in timid and dependent behaviour, and in emotional immaturity.

(3) *Deep Sleep*

Braithwaite (1950), Ström-Olsen (1950) and others consider hypersomnia to be an important aetiological factor in enuresis. EEG studies by Ditman and Blinn (1955) and a carefully controlled experiment by Boyd (1960) failed to confirm this hypothesis.

(4) *Behaviour Disorder*

Several authors, especially Michaels (1955), consider that enuresis is associated with delinquency, both being manifestations of a general behaviour disorder of psychogenic or constitutional origin.

(5) *Emotional Disturbance*

A high incidence of enuresis among children displaying nervous symptoms or emotional disturbance is frequently reported but it is extremely difficult to establish whether in these cases the enuresis is a consequence, a cause, or independent of the emotional difficulties. Certainly increased environmental stress, as in war-time conditions, tends to precipitate the disorder (Burt 1940).

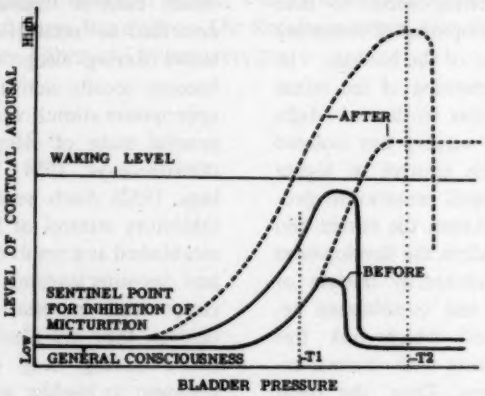


Fig. 1. General consciousness of bladder control during sleep, before and after achievement of continence.

(6) *Specific Dynamic Connotation*

Several writers, particularly those of psychoanalytic persuasion, suggest that enuresis may play a specific role in the psychodynamics of neurotic patients. Sexual or aggressive connotations are most frequently postulated.

Many of these aetiological theories were tested by Hallgren (1956, 1957) in a recent extensive and carefully controlled investigation in Stockholm. His findings support most of the hypotheses described

and he presents evidence in support of his view that, in some enuretics, the condition is primarily genetically determined though the manifestation of the genes is modified by environmental factors.

It seems clear then that nocturnal enuresis may arise from a variety of causes and that it is usually multi-factorial. This is consistent with the developmental theory described in the previous section. The course of development of bladder control would be expected to be affected by abnormalities affecting any one of the interacting functions. Various physical and physiological abnormalities—e.g., spinal deformities, achalasia of the detrusor, abnormalities of urine chemistry, autonomic dysfunctions and neural immaturity might affect the development of the 'sentinel' point, the cortical arousal mechanisms, the waking level and the micturition threshold. The findings of experimental psychology indicate that anxiety and similar emotional states have a disruptive effect on learned behaviour, especially when the latter involves complex discriminations (Jones 1960).

Specific Conditioning Treatment of Enuresis

Despite its apparent efficiency the conditioning method of treatment of enuresis has gained little general acceptance during the twenty or so years since the Mowrers' (1938) original paper. There is, however, a suggestion of a reversal of this trend in recent publications.

It is likely that clinicians are reluctant to adopt this method because it is a highly specific cure for the particular symptom of enuresis and therefore thought likely to lead to 'symptom substitution', the disappearance of the bed-wetting being followed by appearance of some other symptom. Despite evidence to the contrary, this prejudice dies hard and is probably important in the genesis of two very common misapprehensions concerning the nature of the technique. It is frequently said that the method is punitive (and even that the patient is submitted to electric shocks—though this is only true of Crosby's (1950) modification of the method). In fact the patient experiences no more pain than that deriving from the use of an alarum clock and, as the bed remains dry even during the period of training, the treatment is frequently a source of great satisfaction to the patient. It is also argued that conditioning cannot be involved as the ringing of the bell occurs last in the sequence and backward conditioning is extremely difficult to establish. In fact, the bell is not employed as a conditioned stimulus. Owing to the prevalence of these false beliefs it is worth while to consider the rationale of the method in some detail.

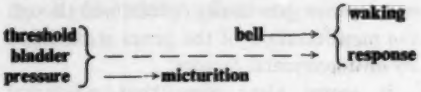
Mowrer and Mowrer (1938) were the first to devise and to make systematic therapeutic use of an efficient apparatus for waking a child immediately after the onset of micturition. They argued that to awaken the child at this time would have the effect of conditioning the waking response to the stimulus of bladder pressure at or near the threshold value for micturition. As waking was accompanied by the inhibition of further urination it was also postulated that this inhibition of urination would become conditioned to bladder pressure approaching the threshold level of urination and would therefore, on future occasions, raise this threshold. They do not state clearly whether they consider that the inhibition of further urination is an involuntary act simultaneous with waking or a voluntary act following waking.

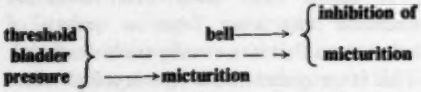
In fact, the response to a waking stimulus such as a bell reciprocally inhibits micturition. Thus the bell, an *unconditioned* stimulus, evokes two simultaneous responses, waking and a reflex inhibition of micturition. If it is arranged that the

bell rings immediately following the onset of micturition, both these responses occur in a temporal relationship to the stimuli evoking micturition appropriate for conditioning to occur. The conditioning process can be schematically represented as follows (UCS = unconditioned stimulus, CS = conditioned stimulus, UCR = unconditioned response, CR = conditioned response):

$$\left.\begin{matrix} UCS_1 \longrightarrow \\ UCS_2 \\ (CS) \end{matrix}\right\} \begin{matrix} UCR_1 \\ \longrightarrow UCR_2 \end{matrix} \left\{\begin{matrix} \\ (CR) \end{matrix}\right.$$

UCS_1 is the original stimulus for response UCR_1. UCS_2 is the original stimulus for response UCR_2, which is usually neglected in conditioning experiments. If UCS_2 consistently precedes UCS_1 by some brief interval of time, a 'connection' is established between UCS_2 and UCR_1, such that UCS_2, if presented alone, will evoke UCR_1. Thus UCS_2, originally the unconditioned stimulus for UCR_2, becomes also the conditioned stimulus for UCR_1. UCR_1, originally the unconditioned response to UCS_1, becomes also the conditioned response to UCS_2. If the stimuli and responses under discussion are substituted for these symbols the following two patterns result.





Thus bladder pressure, in addition to being the unconditioned stimulus for micturition, becomes the conditioned stimulus for the inhibition of micturition and for an independent waking response.

Conditioned responses are normally established to a specific conditioned stimulus such as a tone of a certain pitch and loudness. If, after conditioning, the subjects are tested with tones higher or lower in pitch or of greater or lesser loudness than the one originally used as the conditioned stimulus, the conditioned response is evoked but less strongly than to the original stimulus. In general, the strongest reaction occurs in response to the stimulus originally conditioned, but similar stimuli also evoke a reaction, the strength of which varies with the difference between

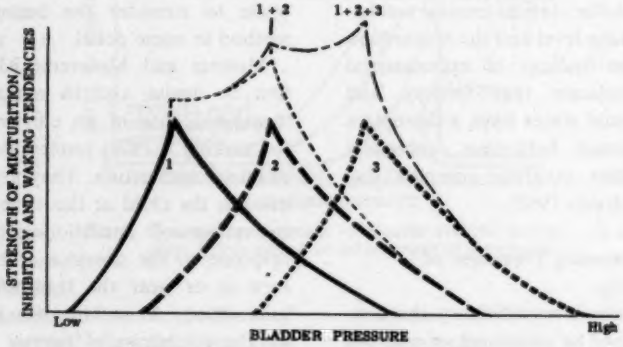


Fig. 2.
Summation of generalisation gradients for conditioned response tendencies:
1 to 3 = individual generalisation gradients
1 + 2 = summation of gradients 1 and 2
1 + 2 + 3 = summation of gradients, 1, 2 and 3.

the original and test stimuli. This phenomenon is known as 'stimulus generalisation' and the gradients of diminishing reaction along stimulus dimensions are referred to as 'generalisation gradients'.

If training occurs in relation to one stimulus value and is then continued at another value the resulting generalisation gradients summate. The successive summations of the gradients relevant to the conditioned responses with which we are concerned are schematically presented in Fig. 2.

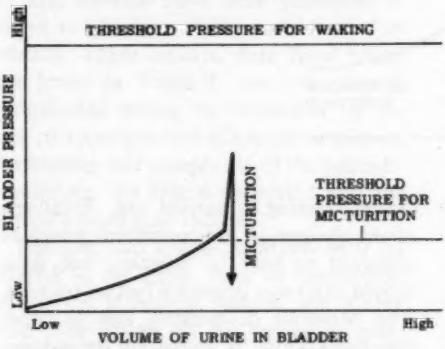


Fig. 3. Nocturnal urination of enuretic before training.

The postulated sequence of events during the treatment of an enuretic by arranging that he is awakened by a bell immediately after the onset of nocturnal urination is schematically portrayed in Figs. 3–6. On the first occasion (Fig. 3) bladder pressure reaches a threshold value for urination well below the threshold for awakening. The child urinates and, as the bell rings, conditioning occurs in the manner described. Thus, on the second occasion (Fig. 4) both the micturition and waking thresholds are distorted by the generalisation gradients of the tendencies conditioned on the first. A somewhat greater volume of urine in the bladder is now tolerated but urination is still induced at a pressure well below the waking threshold and fresh conditioning of the urination—inhibitory and the waking tendencies occurs. On the third occasion (Fig. 5), the thresholds are further distorted by the summated generalisation gradients. Still more urine in the bladder is tolerated but micturition still occurs below the waking threshold and further reinforcement, by the bell, of the conditioned tendencies follows. At this stage, however, there is a range of pressures at which the waking threshold is below that for micturition. At the next stage (Fig. 6) the thresholds are grossly distorted and the range over which the waking threshold is below the micturition threshold is broad.

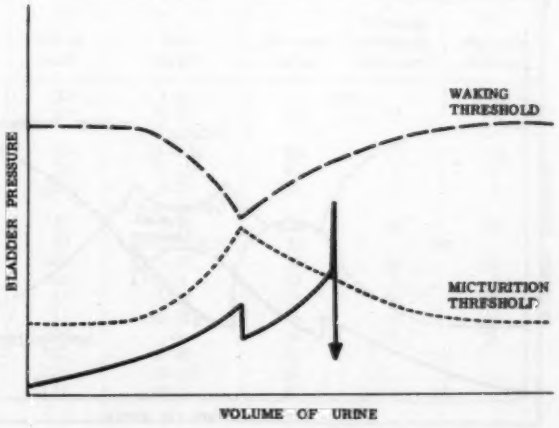


Fig. 4. Nocturnal urination of enuretic during training—stage 1.

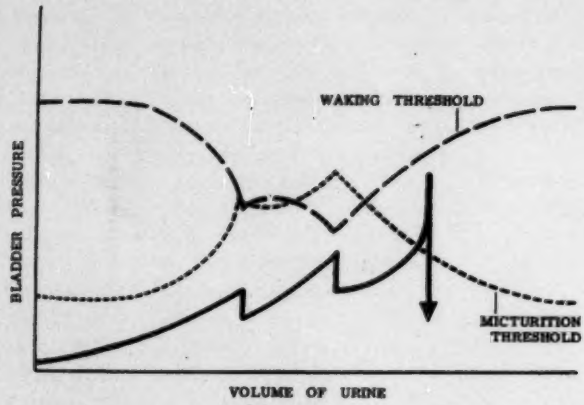


Fig. 5. Nocturnal urination of enuretic during training—stage 2.

The patient now awakens spontaneously at a bladder pressure at which he previously micturated.

Ultimately, most patients develop the ability to sleep through the night without waking to urinate. Thus, the training process enables the child to reach a stage from which normal development can proceed, anatomical growth and the influence of the cortical inhibitory centres on the detrusor muscle tone leading to increased bladder capacity. Apart from these factors, the training would only bring the child to the point at which the strength of the cortical impulses is just sufficient to inhibit urination in the conditions under which training is carried out. Relatively slight changes in the conditions would be expected to lead to 'relapses' but these would in no way prejudice further training. The Mowrers deliberately aim to upset this equilibrium by increasing the evening fluid intake at the stage in training when the child consistently wakes spontaneously to urinate. The increased fluid usually produces further bed-wetting, and, therefore, allows extended training with the bell. When spontaneous waking is achieved in these conditions the apparatus is removed and the intake of fluid is reduced to the normal amount.

Space does not allow a discussion of the

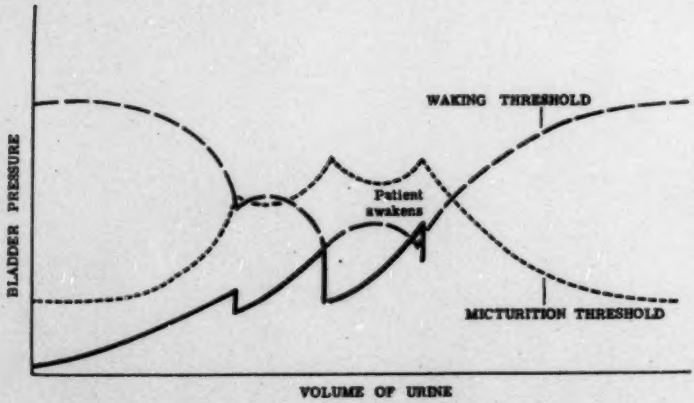


Fig. 6. Nocturnal urination of enuretic during training—stage 3. At asterisk patient awakens.

apparatus and of the practical techniques of application of this method but what is important is that the technique adopted does allow conditioning to occur and allows the conditioned response tendencies to grow to a maximum.

A sufficient number of reports on the treatment of fairly long series of patients by this method have been published to arrive at a fairly realistic evaluation of its efficacy. These reports, data from which are listed in Table I, are not directly comparable owing to variations in the type of apparatus and technique employed, the severity and complexity of the patients' conditions, the follow-up period and the author's criteria of success.

The last two entries, marked with an asterisk in the Table, refer to commercial use of the technique. Martin and Kubly base their figures on the replies to questionnaires issued to those who had purchased the apparatus (in the U.S.A.) over a period of 18 months. Lowe's figures are those of an unqualified practitioner who offers a personal service in his clients' homes.

All authors fail to find evidence of 'symptom substitution' though special attention was paid to this question. The reverse is, indeed, commonly reported, favourable changes of personality and attitude frequently following the remission of this troublesome condition.

These investigations throw little light on the question of the nature of the differences between those who respond to this treatment and those who fail. Lack of co-operation, whether voluntary or involuntary, is undoubtedly a factor in some cases, but this is by no means the only factor. There is little evidence to suggest that 'acquired' enuresis is less responsive than 'primary' enuresis but this is a hypothesis worthy of further investigation.

Though most of the studies listed in the Table did not include control groups it is evident that the degree of success achieved by this method is well in excess of the spontaneous remission rate. It is apparent that, if widely adopted, the specific conditioning method of treatment is capable of significantly reducing the incidence of enuresis nocturna at the later ages of childhood.

TABLE I

Author	*No. of cases*	*Age range*	*Per cent cured*	*Per cent markedly improved*	*Per cent failures*
Mowrer and Mowrer (1938) ..	30	3–13	100		0
Davidson and Douglas (1950) ..	20	5–15 (+ 2 adults)	75	25	0
Crosby (1950)	35	3½–10½	88	3	9
	23	11–28	83	5	12
Sieger (1952)	106	3–15 (+ 4 adults)	89	7	4
Geppert (1953)	42	5–10	74	16	10
Baller and Schalock (1956)	55	Median 9.5	70	30	
Wickes (1958)	100	5–17	50	24	26
Gillison and Skinner (1958) ..	100	3½–21	88	5	7
Freyman (1959)	15	5–14	33	40	27
Murray (1959)	33	3½–18½	75	9	16
*Martin and Kubly (1955)	118	—	56	18	26
*Lowe (1959)	322	5–10	88	12	
	276	10–16	88	12	
	171	16 +	85	15	

SUMMARY

The nature and aetiology of enuresis nocturna is discussed in relation to the normal course of development of voluntary and nocturnal control of micturition. Normal maturation and learning establish cortical 'centres' which inhibit reflex micturition. Ultimately these centres remain active during sleep and nocturnal continence is achieved.

Various physical, physiological and psychological abnormalities have been shown to be associated with enuresis and it is argued that these are of aetiological importance because they disrupt the normal course of development.

The specific conditioning method for the treatment of enuresis, although efficient, has been resisted by clinicians, who are suspicious of symptomatic treatments. Possibly for this reason misapprehensions concerning the nature of the technique are common. The rationale of the method is considered in detail and, in particular, it is shown that the alarum bell does not operate as a conditional stimulus but as an unconditioned stimulus which both wakes the patient and inhibits micturition. During the training, bladder pressure at the threshold for reflex micturition becomes the conditioned stimulus for the inhibition of micturition and for waking. The training process enables the patient to reach a stage from which normal development can proceed.

A brief review of published reports on the efficacy of this treatment shows that the degree of success is well in excess of the spontaneous remission rate. Further research is necessary to establish the nature of the differences between those patients who respond to the treatment and those who fail.

RÉSUMÉ

Traitement de l'énurésie nocturne par le conditionnement spécifique

Discussion de la nature et de l'étiologie de l'énurésie nocturne par rapport au cours normal du développement du contrôle volantaire et nocturne de la miction. La maturation normale et l'éducation instaurent des centres corticaux qui inhibent le reflexe de miction. Ces centres finissent par demeurer actifs pendant le sommeil et la continence nocturne est obtenue.

On a démontré que diverses anomalies physiques, physiologiques et psychologiques sont associées à l'énurésie et on prétend que celle-sci ont une importance étiologique du fait qu'elles rompent le cours du développement.

Bien qu'efficace, la méthode spécifique de conditionnement du traitement de l'énuresie s'est heurtée à la résistance des cliniciens, prévenus contre les traitements symptomatiques. C'est probablement pour cette raison que des méprises relatives à la nature de la technique sont fréquentes. Un exposé raisonné de la méthode est considéré en détail et en particulier il est démontré que la sonnerie du réveil n'agit pas comme un stimulant conditionnel mais comme un stimulant inconditionné qui non seulement réveille le malade mais inhibe la miction. Au cours de l'entraînement, la pression de la vessie au seuil du reflexe de miction devient le stimulant conditionné provoquant l'inhibition de la miction et le reveil. Le processus de l'entraînement permet au malade d'atteindre une étape à partir de laquelle le développement normal peut se poursuivre.

Une brève revue des travaux publiés sur l'efficacité de ce traitement montre que le succès atteint l'emporte de loin sur le nombre des rémissions spontanées. Cependant de nouveaux travaux sont nécessaires pour établir la nature des différences entre malades sensibles à ce traitement et malades qui lui sont réfractaires.

ZUSAMMENFASSUNG

Spezifische bedingende Behandlung der Enuresis nocturna

Das Wesen und die Ätiologie der Enuresis nocturna wird hinsichtlich des normalen Verlaufes der Entwicklung der willkürlichen und nächtlichen Kontrolle des Harnabgangs besprochen. Normale Reifung und Erziehung setzen Rindenzentren ein, die den reflexen Harnabgang hemmen. Schliesslich bleiben diese Zentren aktiv während des Schlafes und die Reinlichkeit bei Nacht ist erlangt.

Es ist gezeigt worden, dass verschiedene physikalische, physiologische und psychologische Abnormitäten mit Enuresis verbunden sind und es wird behauptet, dass diese eine ätiologische Bedeutung besitzen, weil sie den normalen Gang der Entwicklung unterbrechen.

Obgleich die spezifische bedingende Methode wirksam ist, ist sie auf den Widerstand der Kliniker gestossen, denn diese sind gegen symptomatische Behandlung eingenommen. Vielleicht sind aus diesem Grunde die Missverständnisse hinsichtlich des Wesens der Technik so häufig. Eine rationale Darlegung der Methode wird ausführlich betrachtet und es wird, insbesondere, bewiesen, dass das Läuten des Weckers nicht wie ein bedingter Reiz sondern wie ein unbedingter Reiz wirkt, welcher sowohl den Patienten aufweckt als auch den Harnabgang hindert. Während der Übung wird der Blasendruck auf der Schwelle der reflexen Harnentleerung der bedingte Reiz für die Hemmung der Harnentleerung und für das Aufwachen. Der Übungsprozess gibt dem Patienten die Fähigkeit, eine Stufe zu erreichen, von der aus eine normale Entwicklung fortfahren kann.

Eine kurze Übersicht der veröffentlichen Berichte über die Wirksamkeit dieser Therapie zeigt, dass Erfolge weit über die Zahl der spontanen Heilungen hinaus gehen. Fernere Nachforschungen sind nötig, um die Beschaffenheit der Unterschiede zwischen denjenigen Patienten, die auf diese Behandlung reagieren und denjenigen, bei denen sie versiegt, festzusetzen.

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NOTICE

American Films on Public Health Subjects

THE Film Catalogue of the U.S. Department of Health, Education and Welfare contains over 300 entries of films on a wide variety of public health subjects. Their Public Health Service appears to have played a part in the production of all the films and as a result a most valuable collection has been built up. Films on bacteriological and sanitary problems naturally make up the majority of entries and public health laboratory techniques are particularly well covered.

None of the films is immediately concerned with child neurology. If they were, our readers might well be provoked to try to borrow a copy. Readers may recall an earlier article on '*The Movement of Films Between Countries*' (*Cerebral Palsy Bulletin*, Vol. 1, No. 6, 1959, p. 19) which explains some of the intricate formalities that have to be satisfied.

The coverage of subjects in the catalogue looks excellent and it seems most desirable that the free interchange of films of this character should be increased between their countries of origin. The skilled time and enthusiasm that goes into making technical films on medical matters is rarely costed, but all who have taken part will recognise that it is considerable. It is thus a waste of valuable resources when much-needed technical films such as appear in plenty in this catalogue are duplicated in different countries.

There are 2,000 or so medical films available in this country, according to the records of the Scientific Film Association. The majority have been made here but too often they are not available overseas, and likewise expensive film productions exist in other countries and are not known here. To avoid wasteful film making, when skilled resources are not unlimited, there needs to be a greater interchange of information on what is available. The barriers to the exchange of such films must also be diminished. Once sound liaison is established between groups interested in the same topics, any exchange of films by doctors travelling between one country and another could operate very usefully.

S. F. Woodward

The Metabolism of Indole-Compounds in Phenylketonuria

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PHENYLKETONURIA is a hereditary conditions characterised by mental retardation and the presence of phenlypyruvic acid in the urine. Children with this condition appear to be quite normal at birth. Between three and six weeks phenylpyruvic acid can be detected in the urine, and the child fails to develop mentally at the normal rate. In early childhood the children have an attractive appearance, with nearly normal stature and head size. Characteristically, they are blond with blue eyes and fair skin. They tend to suffer from eczema.

Neurologically, phenylketonurics show no paralysis and no changes in muscle tone, though there is accentuation of both superficial and deep tendon reflexes. Electroencephalography shows some abnormal features, such as a long RSP complex with multiple seizure foci, and convulsions sometimes occur. The majority of these patients have an I.Q. of 30 or less, but a few high-grade cases with near-normal intelligence have been described.

Phenylketonuria occurs once in every 40,000 births in the United States and is transmitted by an autosomal recessive gene. The heterozygous carriers of phenylketonuric genes may be identified by means of phenylalanine tolerance tests, in which 0.1 g. per kg. of body-weight of L-phenylalanine is administered by mouth after an overnight fast and plasma phenylalanine levels are determined 1, 2 and 4 hours after the test dose. As a general rule, heterozygous persons have levels twice as high as normal persons.

Phenylketonuria is caused by a deficiency of phenylalanine hydrozylase, the liver enzyme which converts phenylalanine to tyrosine. This in turn causes three kinds of effects:

(1) The excessive phenylalanine is converted, by transaminase, to phenylpyruvic acid. This in turn is converted to phenyl-lactic acid, phenylacetic acid, and phenylacetylglutamines which are excreted in the urine. In addition *o*-hydroxy phenylacetic acid, *m*-hydroxy phenylacetic acid, and indole products derived from tyrosine and tryptophane are also seen in the urine of such patients.

(2) The excessive phenylalanine also inhibits the normal pathways of tyrosine metabolism. There is a decreased production of melanin, and this is responsible for the light pigmentation of the skin and hair of such patients. There is also a disturbance in epinephrine production, since phenylketonurics have unusually low blood-epinephrine levels.

The studies reported here were aided in part by grants from the Illinois Mental Health Fund and the U.S. Public Health Service.

(3) The excessive phenylalanine or one of its products probably causes some, as yet unexplained, damage to the central nervous system; this is characterised by mental retardation, epileptic seizures, and abnormal electroencephalogram changes.

In 1953, Bickel, Garrard and Hickmans[1], at the suggestion of Dr. L. I. Woolf, prepared a special protein hydrolysate low in phenylalanine content. When this was administered to a phenylketonuric child the phenylalanine level in the plasma was gradually reduced, and the phenylpyruvic acid disappeared from the urine. Recently, a number of reports have appeared assessing the true value of this diet in bringing about normal development in phenylketonuria[2,3,4]. There is general agreement that the diet may be of considerable value in the very young infant, but it becomes increasingly less effective in the child started at a later age, and it is probably of no value in older children and adults.

The cause of the mental defect in phenylketonuria is unknown. Despite extensive studies both *in vivo* and *in vitro* there is still no clear evidence that any of the phenolic derivatives of phenylalanine, or phenylalanine in itself, acts as a 'toxic' substance on the brain[5]. For this reason, it might be worth while to speculate on the possible role of indole compounds in the mental defect of phenylketonurics.

Nature of the Indole Disturbance in Phenylketonuria

A simplified scheme for the normal degradation of tryptophane in mammalian systems is given in Fig. 1. The conversion of tryptophane to 5-hydroxy tryptophane in the first step is as yet poorly understood and defined. Udenfriend and his co-workers[6] have shown that when they administered DL-tryptophane-2-C^{14} the label could be recovered in 5-hydroxy tryptophane in toad venom gland, suggesting that there might be a direct pathway. The conversion of 5-hydroxy tryptophane to 5-hydroxy tryptamine in the second step has been extensively studied.

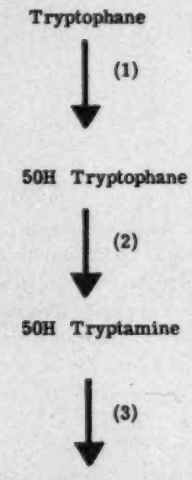

Fig. 1. Normal pathway for the degradation of tryptophane.

The reaction is catalysed by 5-hydroxy tryptophane decarboxylase[7] and requires pyridoxal phosphate as a cofactor[8]. The 5-hydroxy tryptamine is then converted to 5-hydroxy indoleacetic acid in the third step and excreted in the urine. This reaction is catalysed by momoamine oxidase.[9, 10]

In 1954, Armstrong and Robinson[11] showed that indoleacetic acid and indolelactic acid are excreted in amounts of 0.02 to 0.15 grammes per day or less in patients with phenylketonuria. This finding has been confirmed by Jepson[12] and by Ferrari and his co-workers.[13]

More recently, Pare, Sandler and Stacey[14, 15, 16] have demonstrated a defect of 5-hydroxy indole metabolism in phenyl-

ketonuria. This is characterised by a decrease of 5-hydroxy indoleacetic acid from 7.2 mg. per g. creatine in 32 controls to 2.2 mg. per g. creatine in 49 phenylketonurics. Similar results were found in serum 5-hydroxy tryptamine, where the

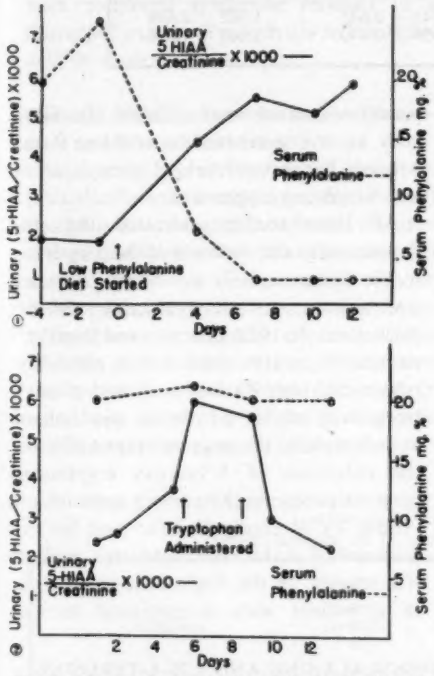

Fig. 2. Changes in serum phenylalanine levels and urinary 5-hydroxy indoleacetic acid excretion during transition to a low-phenylalanine diet. (From Baldridge, R. C. *et al. Proc. Soc. exp. Biol. Med.* 1959, **100**, 529).

mean level in 49 phenylketonurics was 71.2 ng./ml. compared with 283 ng./ml. in 31 controls. When low phenylalanine diets were administered there was a significant rise in the mean serum 5-hydroxy tryptamine levels and in the excretion of 5-hydroxy indoleacetic in the urine. Essentially the same results have been reported by Baldridge and his co-workers[17] and Berendes *et al*[18] (Fig. 2). We have lately demonstrated similar changes with 3.4 dihydroxy phenylalanine (DOPA). The levels of nor-epinephrine and epinephrine were found to be markedly decreased in both the plasma and urine of untreated phenylketonurics as compared with normal controls. These changes were reversible by the use of a low phenylalanine diet in phenylketonurics[28]. Since the tyrosine levels remain unaltered, these changes could be attributed to the inhibition of DOPA decarboxylase by phenylalanine metabolites *in vivo*, which confirms the previous impression that there is similarity in the behaviour of the pyridoxal phosphate dependent amino-acid decarboxylases. A similar depression has recently been observed among the heterozygous carriers for phenylketonuria in our laboratory[19]. A group of parents of phenylketonuric children and comparable normal controls were given 0.1 g. per kg. bodyweight of L-phenylalanine by mouth. Urine was collected during the first six hours after the initial load and again for an additional twelve hours. The mean excretion of 5-hydroxy indoleacetic acid during the first six hours was 0.959 mg. among the 15 heterozygotes, compared with 1.543 mg. among the 16 normal controls. The difference could no longer be demonstrated during the next twelve-hour period (Table I).

Aetiology of the Indole Disturbance in Phenylketonuria

In the beginning, some workers[20] held the view that the abnormality of indole metabolism in phenylketonuria might represent a separate genetically determined metabolic defect. This viewpoint is no longer held for a number of reasons. In the first place, the decrease of 5-hydroxy tryptamine in the blood and of 5-hydroxy indoleacetic acid in the urine is readily reversible in the phenylketonuric patient when phenylalanine is withheld from the diet. In the second place, the decrease of 5-hydroxy indoleacetic acid in the urine

TABLE I—URINARY EXCRETION OF 5-HYDROXY INDOLEACETIC ACID IN THE URINE AFTER THE ADMINSTRATION OF 0.1 g./kg. BODY-WEIGHT OF L-PHENYLALANINE BY MOUTH

	Urinary 5-hydroxy indoleacetic acid (mg./volume)	
	0–6 hours	*6–18 hours*
16 Controls	1.543 ± 0.645	1.902 ± 0.648
15 Heterozygotes	0.959 ± 0.470	1.869 ± 1.090

was noted in the heterozygote only during the first six hours after the phenylalanine load when the plasma phenylalanine level is at its peak. However, the definitive demonstration of the fact that this is a secondary manifestation due to excessive phenylalanine has been undertaken in normal rats made phenylketonuric by means of added phenylalanine and tyrosine, as described by Auerbach and his co-workers[21]. In these experiments[22], it was found that the urinary excretion of 5-hydroxy indoleacetic acid decreased as the urinary phenylpyruvate increased as a result of the added phenylalanine and tyrosine in the diet. When the supplementation was discontinued and the phenylpyruvate excretion decreased the urinary 5-hydroxy indoleacetic acid rose up to the previous normal levels (Table II). Similarly, an inverse correlation could be found between the serum-levels of phenylalanine and 5-hydroxy tryptamine.

All these studies indicated that the depression in the synthesis of the 5-hydroxy indole compounds is secondary to excessive amounts of phenylalanine and its derivatives. In 1958, Davison and Sandler[13] showed by *in vitro* studies that phenylpyruvic acid, phenylacetic acid, and phenyllactic acid inhibit 5-hydroxy tryptophane decarboxylase, the enzyme responsible for the reduction of 5-hydroxy tryptamine from its precursor 5-hydroxy tryptophane (Table V). Recently Sandler and his co-workers[24 25] have confirmed this with *in vivo* studies. In the first study, carried out in a patient with a carcinoid tumour

TABLE II—EFFECT OF 3.75 PER CENT L-PHENYLALANINE AND 3.75 L-TYROSINE ON THE EXCRETION OF PHENYLPYRUVATE AND 5-HYDROXY INDOLEACETIC ACID IN THE URINE

Age (weeks)	*Phenylpyruvate (mg./24 hr.)*		*5-hydroxy indoleacetic acid (kg./24 hr.)*	
	Controls	*Experimental*	*Controls*	*Experimental*
3½	0.74	7.43	27.5	2.4
5	0.36	1.60	26.2	7.7
7	0.42	5.45	25.4	14.1
7½	0.08	3.39	22.2	16.6
9	0.30	6.58	21.2	19.2
10	0.20	4.40	22.2	8.9
11	0.33	2.12	50.3	15.0
12	0.74	2.55	30.2	14.7
13	0.43	2.02	32.6	14.0
14	0.72	0.84	23.0	15.7
15	0.98	1.37*	21.4	38.3*
16	1.13	2.11*	32.4	40.2*
17	0.84	0.62*	24.2	13.2*

* Returned to stock diet.

secreting 5-hydroxy tryptophane, the administration of phenylacetic acid appeared to decrease the urinary output of 5-hydroxy tryptamine and 5-hydroxy indoleacetic acid. In the second study, the administration of phenylacetic acid to five patients with carcinoid syndrome resulted in a decreased output of 5-hydroxy indoleacetic acid in three of them.

Possible Relation Between Indole Disturbance and Mental Deficiency

The aetiology of the mental defect in phenylketonuria is unknown. However, it has been shown that if the affected children are treated with a low phenylalanine diet at an early age near-normal mental development will result. A similar course of treatment in an older affected child is of little or no value. This indicates that the beneficial effect of the diet must in some way be related to the development of the brain during early infancy.

During the past year, we[26] have reported on the development of 5-hydroxy tryptophane decarboxylase activity in the newborn rat kidney. The data, which are summarised in Table III, show that this enzyme is absent in the foetal and newborn rat, and the activity of the enzyme increases gradually, reaching an adult level by the age of 33 days. One could speculate that the excessive amounts of phenylalanine in the phenylketonuric child would further inhibit an already immature enzyme system, and this in turn will result in a decrease of 5-hydroxy tryptamine, which is essential to the developing brain. Studies to elucidate this process are now in progress.

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TABLE III—INHIBITION OF 5-HYDROXYTRYPTOPHANE DECARBOXYLASE BY PHENYLALANINE METABOLITES
(From Davidson, A. N. and Sandler, M. *Nature* 1958, **181**, 186.

Additions	*Conc. (umoles/ 3 ml.)*	*Output of CO^2 (ml./40 min.)*	*Inhibition (per cent)*	*Inhibition of dihydroxy-phenylalanine decarboxylase (per cent)*
Control	—	75.7	—	—
Phenylpyruvic acid ..	100	15.7	79.3	100
	50	38.3	49.5	100
	10	74.2	2	77
Phenyl-lactic acid	100	36	52.5	100
	50	66	13	77
	10	71.2	6	50
Phenylacetic acid	100	12.5	83.5	50
	50	24.2	68	0
	10	—	—	0
L-Phenylalanine	100	65.7	13	0

Guineapig kidneys were homogenised in ice-cold M/30 Sorensen's phosphate buffer pH 6.8. The homogenate was centrifuged in the cold for 5 min. and 2 ml. of the supernatant (equivalent to 2 g. of original wet tissue) used in each Warburg flask. 25 ug. of pyridoxal phosphate and the inhibitor dissolved in phosphate buffer, was added. The volume was adjusted to 2.75 ml. with phosphate buffer. After gassing for 2 min. with nitrogen, the flasks were equilibrated at 37° for 10 min. and 0.25 ml. of DL-5-hydroxy-tryptophane (10 umoles) added from the side-arm. Controls and blanks were also used. Carbon dioxide output was determined for 40 min.

TABLE IV—KIDNEY 5-HYDROXYTRYPTOPHANE DECARBOXYLASE ACTIVITY IN RATS OF DIFFERENT AGES

Age	*No. of exp.*	*No. of animals*	*5-hydroxytryptophane decarboxylase activity**
Foetal + ...	3	45	0
1 day	3	23	0
2–5 days	4	24	0.21 ± 0.21
7–12 days	5	26	0.38 ± 0.10
21–23 days	7	21	0.66 ± 0.13
33 days	4	8	1.19 ± 0.47
Adults	14	14	1.34 ± 0.58

* Mean values expressed as umoles 5-hydroxtryptamine formed per 100 mg. of dry weight of kidney per hr.
+ 1–3 days before term.

SUMMARY

Considerable efforts have been directed towards the elucidation of the phenol compounds in the pathogenesis of phenylketonuria, but relatively little has been done towards defining the role of the indole compounds.

Observations have been made regarding the levels of 5-hydroxy tryptamine, 5-hydroxy indoleacetic acid, and 5-hydroxy tryptophane decarboxylase in phenylketonuria, and their possible role in causing mental deficiency in the developing infant is discussed.

RÉSUMÉ

Métabolisme des composés de l'indole dans la phénylcétonurie

On a fait des efforts considérables pour élucider la question des composés du phénol dans la pathogénie de la phénylcétonurie mais on n'a relativement pas fait grand chose pour préciser le rôle des composés de l'indole.

Nous trouvons ici des observations relatives aux taux de la 5-hydroxy tryptamine, de l'acide 5-hydroxy indoleacétique et de la 5-hydroxy tryptophane décarboxylase dans la phénylcétonurie et une discussion de leur rôle éventuel dans les causes de déficience mentale chez les enfants en cours de croissance.

ZUSAMMENFASSUNG

Stoffwechsel der Indolverbindungen bei Phenylketonuria

Erhebliche Bemühungen sind auf die Aufklärung der Rolle der Phenolverbindungen in der Pathogenie der Phenylketonuria gelenkt worden aber es ist verhältnismässig wenig getan worden, um die Rolle der Indolverbindungen zu erklären.

Hier findet man Beobachtungen hinsichtlich des Gehalts an 5-Hydroxytryptamin, 5-hydroxyindol-essigsäure und 5-Hydroxytryptophan decarboxylase bei Phenylketonuria und Erörterung über ihre eventuelle Rolle in den Ursachen des Schwaschsinns bei aufwachsenden Kindern.

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Phenylketonuria: Clinical Aspects

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THE phenylketonuric infant does not look any different from others at birth. Unlike cases of mongolism or Hurler's disease he has no characteristic physical features by which he can be recognised. He usually thrives quite well for a time and it may well be not until he is a year old, and possibly much later, that the parents realise there is anything amiss. In Berg's series (see below) 3 cases were not diagnosed until the age of 4–5 years. It is for this reason that the chemical aspects of this metabolic disorder are so important, especially as it is now suggested that specific treatment with an appropriate diet at a very early age will prevent damage to the brain, at least in some cases.

The clinical physical signs are not really helpful in the diagnosis. I am not convinced that the tendon reflexes are particularly exaggerated. As to the dilution of pigment, this is a real phenomenon (Cowie and Penrose 1951, Berg and Stern 1958) but it is not much help in a particular case. One of our phenylketonuric children in the study reported by Berg and Stern had a dark iris, 6 had a medium and 20 a light one. In family no. 1 both normal siblings happened to have lighter irises than the patient. Similarly, in a dark family, the hair of the affected child will still be dark though probably fairer than that of the normal children.

Early Diagnosis

There is thus a real doctor's dilemma! On the one hand early diagnosis is essential if the child is to be normal; on the other hand there are no reliable physical signs and diagnosis rests on a test of the urine. The only method of discovering early cases would seem to be routine testing of infants. But this would mean testing 40,000 urines to find one positive—a formidable labour! Fortunately this situation only arises with the first case in the family. Once it is known that a relative is affected, especially if it is another sibling, then the arrival of the new baby is awaited with special interest and his urine will no doubt be tested at the end of three weeks and again at six weeks. Urine testing is now routine in mental deficiency hospitals, and local authorities are beginning to extend it to special training centres and schools for the educationally subnormal where the expectation of finding positive results is much greater than in the general public. At the same time the use of previously prepared impregnated sticks ('Phenistix') makes routine testing easier and simpler. They can be used routinely in maternity work and are suitable for application to the wet napkin, which is a great advantage. Gibbs and Woolf (1959) report that false positives are not achieved by this method so that a strong positive result with Phenistix at 3 weeks can be taken as definitive, while any weak reactors should be examined by chromatography.

Progress in the identification of carriers by the methods described by Hsia or otherwise is likely to be rapid in the near future. This will be a formidable task if, as seems reasonable to suppose, these number

1 per 100 of the population. However, this work will also enable the birth of affected children to be anticipated, and appropriate treatment to be applied at an early stage. When assessing the value of routine testing of urine it is perhaps relevant to ask how much use is made of certain observations which are now standard, as for example routine measurement of the specific gravity. The addition of the simple Phenistix test to the routine procedure would perhaps not be unreasonable when the matter is viewed in this light.

Progress of Untreated Case

Just as phenylketonuria has no special physical features, so it is not characterised by any special psychological aspects except a tendency to marked retardation ranging from severe idiocy to near normality in the exceptional case. A typical untreated case at the age of 6 years may have reached the level of a child of 2 years and may only now be beginning to talk and be clean in toilet habits. He may not yet feed himself independently. The condition is, however, not progressive at this stage and the child will continue to develop gradually in mental ability until the age of some 16 years. Just as with other children of imbecile level, so he also can be taught with the exercise of great patience, and he may well be able to attend a special training centre daily from his home under the auspices of the Local Health Authority if he is not in hospital. Fortunately, physical disabilities are uncommon in these cases and the affected children do not usually suffer from cerebral palsy, deafness or blindness. Epilepsy is a common complication. In a series of 800 admissions to the Fountain Hospital, Berg (to be published) noted 11 cases of phenylketonuria (1.38 per cent). One of these cases had epilepsy after admission to hospital but in 4 other cases there was a history of fits, mainly minor attacks, in early childhood. Reduction in size and weight was not a conspicuous feature in this group of cases, but we have found occasional phenylketonuric children with heads so small that they could be described as microcephalic (Brandon *et al.* 1959). This is unusual and suggests that in very severe cases of the condition growth of the brain may be slowed very much after birth. As I have mentioned, most of the patients are indistinguishable from normal children in physical appearance.

The skin in phenylketonuria is delicate. Affected children usually show well-marked dermatographia and there is a tendency to recurrent eczema.

Digital mannerisms have been mentioned (Cowie 1951) as a feature of the behaviour of phenylketonuric children, but these occur only in the lower grade cases, usually of idiot level, and they are by no means confined to this condition, being a common feature of the activity of idiots. They consist of repetitive finger and hand movements, sometimes like the 'hand regard' of the infant in which satisfaction seems to be derived from close observation of finger movements. In other cases there is persistent twiddling of a bootlace or spinning of some object.

On the whole, phenylketonuric patients are robust by comparison with other severely subnormal children. Their expectation of life at birth is somewhat reduced by comparison with normal infants but they are much more viable than children with mongolism or gargoylism. This is a fact to be borne in mind by the family doctor and parents when making plans for the future of the child. The mother may be well able and willing to manage the infant at home, however handicapped, but an older patient of idiot level may be too much for her, especially when the mother herself is older and less capable of carrying the additional burden.

Treatment

Treatment should be started at the earliest possible moment. The only method which can be recommended at present is the reduced phenylalanine diet. This should be commenced as soon as the diagnosis is established. There is probably little point in starting such treatment after the third or fourth year. It is also probable that the need for a strict diet is not so important in older children, though further work is needed on this problem. A primary difficulty with the diet originally was its unpalatability, but this obstacle now seems to have been largely overcome. Two other points need to be watched. In depriving the child of almost all natural proteins he should not be left without any source of phenylalanine; a minimal quantity is essential. Also, the highly artificial diet needs adequate supplements of vitamins and other essential food factors. Woolf *et al.* (1955, 1958) recommend 60 to 120 ml. of cow's milk for children over 18 months of age and somewhat higher figures for younger infants. They set out the detailed composition of the artificial 'milk' used as the main source of protein and also of the daily intake of supplements. The preparation is now commercially available as 'Minafen' (Trufood) or 'Cymogran' (Allen and Hanburys) in Britain, and as 'Lofenalac' (Mead Johnson) or 'Ketonil' (Merck, Sharp & Dohme) in the U.S.A. Any food containing considerable amounts of natural protein is excluded from the diet except the set amount of milk mentioned above. Items which are relatively protein-free, such as kosher margarine, tomato-juice, cornflour, honey, jam, carrots, cabbage, and gluten-free wheat starch, can be used freely. Potatoes are given to older children in controlled amounts. Ordinary white or brown flour is prohibited.

If the diagnosis is made in a child living at home the best course is to arrange for stabilisation on the diet to be carried out in hospital in a paediatric department. The diet can then be continued at home. A booklet entitled *Diets for Sick Children*, published by the Hospital for Sick Children, Great Ormond Street, contains suitable recipes. The basic items of the special diet can be obtained through the National Health Service. Adherence to the diet will need considerable persistance on the part of the mother, and even in hospital will need continual supervision to avoid errors or the stealing by older children of forbidden items from the other children in the ward. Urine testing provides a rough guide to the amount of phenylpyruvic acid being excreted, but blood-phenylalanine measurement by the micromethod is necessary for reliable control.

Advice to Parents

The attitude of the parents to the discovery that their child is handicapped in this way will depend somewhat on whether they already have normal children. If they have they will find it easier to bear the misfortune. The risk of a subsequent sibling being affected is high—one in four—so that most parents would not wish to have further children when informed of this risk.

As to the care of the affected child, he is best looked after at home as an infant, apart from any necessary periods in hospitals for dietary purposes or investigation. It is important that the mother should keep close contact with him during such absences. Daily visits are desirable if possible. If the diet and other treatments are to have the best effect then it is desirable that there should be the psychological stimulus which contact with the mother and the home environment will provide. In cases that do badly, institutional care may be recommended later but this should seldom be necessary before the

age of four or five years and should be regarded as a final resort. Daily attendance at special school or training centre is preferable, but much will depend on the home circumstances and particularly on the parental attitude.

Future Developments

Dietetic treatment is tedious and difficult. In view of lack of controls, validation of results is still *sub judice*. The ideal treatment would be substitution of the missing enzyme. Short of this we (Pare, Sandler, Stacey and Kirman, unpublished observations) have made some tentative trials of possible treatments in connection with the known deficiency of 5-hydroxytryptamine (5HT, serotonin) in this disease. Some patients were given 5-hydroxytryptophane, the precursor of 5HT, in the hope of increasing the amount available. In another trial—iproniazid was given with the object of reducing the destruction of 5HT. In neither case was the mental state of the patients notably improved but this was not anticipated since they were over the age when a response would be expected. On the other hand, biochemical changes in blood and urine were in the anticipated direction and suggest that both of these approaches might be followed further.

SUMMARY

There are no reliable physical signs in phenylketonuria and diagnosis is based on chemical tests. Cases can be readily identified in affected families. All backward children should be tested, and with 'Phenistix' routine testing of babies at 3 weeks is feasible. Carriers are identifiable. The disease is not progressive when established. There are few complications apart from occasional epilepsy and eczema. The expectation of life is better than in mongolism. Treatment with a low phenylalanine diet should be tried at the earliest possible moment, though the results obtained require further validation. Other methods of therapy should be sought. Some patients may attend school or a special training centre from home, while others need hospital care.

RÉSUMÉ

La phénylacétonurie: aspects cliniques

Il n'y aucun a signe physique certain dans la phénylacetonurie. Le diagnostic ne repose que sur les examens chimiques. Il est facilie de déceler les malades dans les familles qui en sont atteintes. Tous les enfants retardés devraient être examinés et le 'Phenistix' permet de faire les examens courants des petits enfants de 3 semaines. Les porteurs sont identifiables. La maladie se stabilise dès qu'elle est établie. Les complications sont rares en dehors de cas sporadiques d'épilepsie et d'eczéma. Les chances de suivie sont plus favorables que dans le mongolisme. Le traitement par régime pauvre en phénylalanine devrait être essayé le plus tôt possible bien que les résultats obtenus réclament de nouvelles confirmations. Il est souhaitable de rechercher d'autres méthodes thérapeutiques. Certains malades vivant dans leur foyer peuvent aller à l'école ou dans les centres de formation spécialisés. D'autres par contre relèvent de l'hôpital.

ZUSAMMENFASSUNG

Phenylketonuria: klinische Aussichten

Es gibt kein sicheres physisches Zeichen bei Phenylketonuria und die Diagnose stützt sich auf chemische Proben. Die Fälle können leicht in befallenen Familien festgestellt werden. Alle zurückgebliebenen Kinder müssten untersucht werden Routineuntersuch-

ungen bei Säuglingen von 3 Wochen ausführbar. Träger sind identifizierbar. Die Krankheit schreitet nicht mehr fort wenn sie festgesetzt worden ist. Komplikationen sind selten ausser gelegentlichen Epilepsia und Exemfällen. Die Lebenswahrscheinlichkeit ist besser als bei Mongolismus Behandlung durch Phenylalaninarme Diät müsstso früh wie möglich versucht werden, obgleich die erlangten Resultate weitere Bestätig ungen erfordern. Andere Behandlungsmethoden müssten gesucht werden. Einige Patienten können daheim die Schule oder spezielle Ausbildungsanstalten besuchen, während andere Hospitalpflege brauchen.

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Tests for Phenylketonuria

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The Importance of Testing

THE development of a rational and effective treatment for one form of mental deficiency, phenylketonuria, made its diagnosis a matter of the first importance. The further finding that some irreversible brain damage occurred in most phenylketonurics during the first few weeks or months of life made it urgently necessary to make the diagnosis in apparently normal babies during the neonatal period, so that treatment could be started; by the time mental retardation has become manifest it is too late for treatment to be fully effective.

It is difficult to diagnose phenylketonuria on purely clinical grounds. During the first few months of life the phenylketonuric resembles any normal baby; during later childhood the untreated phenylketonuric resembles any other grossly mentally retarded child. Fortunately there are extremely simple and reliable tests for phenylketonuria, based on the detection of phenylpyruvic acid in the urine.

How to Test

(a) *With 'Phenistix'*

'Phenistix' [Ames Co. (London) Ltd., Nuffield House, Piccadilly, London, W.1] are strips of heavy absorbent paper, impregnated at one end with ferric ammonium sulphate, a buffer (cyclohexylsulphamic acid) pH 2.3, and a magnesium salt. The impregnated end is dipped briefly (for not more than one second) into the urine, and, if phenylpyruvic acid is present, turns green within a minute, the colour fading only slowly. The test is, for all practical purposes, completely specific and sufficiently sensitive for diagnostic use. If, however, the patient is taking chlorpromazine, there is a risk of a false positive reaction, and if the patient has taken aspirin or salicylates the Phenistix strip turns purple and it would be impossible to see any green colour. The urine must be fresh, or, if more than one hour or so old, must be preserved (preferably with chlorbutol or chloroform) and kept refrigerated. The urine must be free from faecal or other contamination and non-alkaline (see below). It is preferable to avoid early morning specimens, since the concentration of phenylpyruvic acid is lowest at this time.

If a liquid specimen of urine cannot be obtained, the Phenistix can be pressed against the wet napkin. It is necessary to ensure that the napkin is wet, not merely moist, and that the Phenistix is thoroughly wetted. It is best to place the Phenistix strip between two layers of the wettest part of the napkin and to press firmly for half a minute.

If the napkin is only moist and will not wet the Phenistix it is possible to proceed as follows: the Phenistix is dipped into water for about one second and then pressed very firmly against the moistest part of the napkin. The salts in the Phenistix dissolve and print on to the napkin, rather like a rubber stamp; in the presence of phenylpyruvic acid a green patch the size and shape of the impregnated end of the Phenistix appears on the napkin. This modification is, however, less sensitive and less specific than the conventional technique. *Para*-hydroxyphenyl-

pyruvic acid, found in the urine of 1 per cent of babies, gives only a very fleeting green colour, impossible to confuse with phenylpyruvic acid, when Phenistix is used in the conventional way; but by the 'printing' method the two acids are almost indistinguishable. This is presumably due to the different ratios of reagent to chromogen under the two sets of conditions.

(b) *With Ferric Chloride*

Fölling's original test—the addition of ferric chloride to the urine, with the development of a green colour if phenylpyruvic acid is present—is still the cheapest and probably the most widely used. The reagent is a 5 per cent or 10 per cent solution of ferric chloride in distilled water; it keeps for two or three months before throwing down a precipitate and becoming useless. The ferric chloride solution should be added drop by drop to a little urine, shaking after each addition, till a definite colour appears or till there has been added to the urine about half its volume of the 5 per cent solution or one quarter its volume of the 10 per cent solution. The green colour takes a little time, up to five minutes, to reach maximum intensity and then slowly fades, sometimes in minutes, sometimes overnight. The colour varies from a clear green to a deep slate blue, almost black, depending on the concentration of the urine, the presence of phosphates, etc.

The urine must be reasonably fresh, free from faecal or other gross contamination and non-alkaline. A physiologically alkaline urine can be tested if it is very fresh, but phenylpyruvic acid is unstable in alkaline solution. If these precautions are not observed it is possible to get a false negative reaction. If urine has to be stored for some hours or longer, it should have a preservative added and be refrigerated.

Some textbooks recommend acidifying the urine before adding the ferric chloride, but, unless one has a pH meter and can bring the urine to pH 2.3, this is at best useless and at worst nullifies the result. Below pH 2.3 the intensity of the colour given by phenylpyruvic acid and ferric chloride decreases rapidly, but it is very easy to achieve pH values well below 2 by adding dilute sulphuric or hydrochloric acid to the urine, resulting in little or no colour production; a number of cases have been missed on first testing because the urine was acidified. On the other hand, acetic acid is a weaker acid than the ferric chloride solution itself and the addition of acetic acid to the urine before testing is pointless unless it is to act as a preservative. Owing to hydrolysis the ferric chloride solution (pH about 2) is sufficiently acid to bring any but very alkaline urines to near the optimum pH for colour development. Very alkaline urines should be discarded—a negative result obtained on such a urine is meaningless and it is useless to neutralise or acidify a very alkaline urine before testing, since any phenylpyruvic acid originally present will have been destroyed.

The greatest disadvantage of the ferric chloride method is that it requires a liquid specimen of urine and this is often difficult to get. Gibbs and Woolf (1959) found that only 25 per cent of mothers were successful in bringing a specimen of their babies' urine to the welfare centre; on the other hand, when testing with Phenistix on the napkin was substituted, more than 97 per cent of babies could be tested (Gibbs, personal communication). It is possible to drop the ferric chloride solution directly on to the napkin, but a positive reaction is sometimes difficult to see and the reagent rots and stains the cloth.

A number of substances besides phenylpyruvic acid give green colours with ferric chloride. The most important of these is *p*-hydroxyphenylpyruvic acid which has been found in the urine of over 1 per cent

of babies tested. It is difficult to distinguish these two acids using ferric chloride; fortunately Phenistix reacts differently towards them.

(c) *Using Filter paper*

Berry *et al* (1958) instruct the mother to place a piece of ordinary filter paper in the napkin. After wetting, the filter paper is dried without artificial heat and sent through the post to a central laboratory. A number of tests for various conditions can be carried out on the paper. A disadvantage, in this country, is that the available laboratories would find it difficult to cope with specimens from 600,000 babies a year. It is, however, a convenient way of getting a specimen for confirmatory tests if the first test appears to be positive, or if there is a special reason for testing.

(d) *With* 2 : 4-*dinitrophenylhydrazine*

A solution of 2 : 4-dinitrophenylhydrazine in hydrochloric acid gives a yellow precipitate with phenylketonuric urine. A liquid specimen is necessary and other keto-compounds give similar precipitates, but nevertheless this is a useful confirmatory test; only phenylpyruvic acid and *p*-hydroxyphenylpyruvic acid, among the substances commonly found in urine, are positive both by this test and with ferric chloride.

(e) *Confirmatory Tests*

A positive result should never be accepted as conclusive without confirmation. Confirmatory tests are, necessarily, a matter for the laboratory. The best test is probably paper chromatography of blood amino-acids, but paper chromatography of urinary amino-acids or phenolic acids, or quantitative estimation of serum phenylalanine is satisfactory. In phenylketonuria the phenylalanine concentration in the blood and urine rises to many times normal and *o*-hydroxyphenylacetic acid appears in the urine.

It is, naturally, of the first importance that suspected cases should be confirmed with the least possible delay.

The action to be taken while waiting for the result of the confirmatory test must depend on circumstances. In the case of a 'high-risk' infant (see below) giving a positive test for phenylketonuria, it is probably best to start treatment at once. This is, however, not without danger, since a normal infant will go downhill rapidly on a low-phenylalanine diet and it is probably safer not to institute this treatment in the general case on the basis of a single positive test.

Whom to Test

All newborn infants and all mental defectives should have their urine tested for phenylpyruvic acid. Newborn infants in the 'high-risk' group should be tested more intensively than others. The test is so simple and quick that one is amply justified in applying it to all newborn infants, in view of the grave permanent mental impairment that results if phenylketonuria is not diagnosed and treated early. This is in spite of the fact that only one baby in twenty thousand is born with the condition. (This is the best estimate available for Great Britain as a whole; there is evidence that the incidence is much higher in some parts of the country.)

There is a special 'high-risk' group of infants who have sibs known to be suffering from phenylketonuria. Since this is a recessively inherited condition, there is a one in four chance of any sib of an affected individual having the disease himself. So high a probability justifies very careful investigation in this small group. It is advisable to test the urine for phenylpyruvic acid daily from birth onwards for some weeks and to examine the blood-phenylalanine level, preferably by paper chromatography, once a week for the first three weeks of life before accepting a negative finding.

All children suspected of mental retardation should have their urine examined for phenylpyruvic acid. Apart from the obvious desirability of making a diagnosis, there is the possibility that dietary treatment may, to some extent, help children who have not suffered too much brain damage, and it is possible to give the family more accurate genetic counselling than if the test is omitted.

There are large gaps in our knowledge of the regional incidence of phenylketonuria. If all mental defectives, institutionalised or not, were tested as a research project, it would help to close these gaps.

When to Test

Ideally the urine of all infants should be tested when they are two weeks old and again when they are six weeks old. If only one test is possible, this should be done when the baby is three or four weeks old.

The reason for choosing these times is that during pregnancy there is free exchange of amino-acids across the placenta and the maternal liver can metabolise phenylalanine normally both for mother and foetus. The child is therefore born with a normal blood-phenylalanine concentration, and it takes time, in the phenylketonuric, for this concentration to rise to the level where phenylpyruvic acid is excreted in the urine. The time needed varies from about a week to over five weeks in one case. However, there is evidence that irreparable damage of moderate severity can be done if treatment on the low-phenylalanine diet is delayed until the infant is five or six weeks old. It therefore seems best to test at two weeks, by which time most phenylketonuric infants will be excreting phenylpyruvic acid but brain damage should still be undetectable, and again at six weeks to pick up the few late starters. As a compromise one may test once only at the age of three or four weeks, thereby missing only a very small proportion of the late starters but still keeping brain damage slight, while some authorities prefer to test once at six weeks so that they miss no positive cases. It is worth emphasising that the damage done to the brain during these first weeks of life is never detectable at the time but shows itself much later in a lowered I.Q.

The question of who is to carry out the test will be answered differently in different areas. Where a health visitor sees the fortnight-old baby in its home, this is the ideal opportunity for testing, but where the baby is tested at the welfare centre, the first test will have to wait till the child's first visit. The midwife does not usually see the baby after a fortnight and so, although she could carry out the first test at that time, a second test at about six weeks would have to be done by someone else. In any case it is very desirable that the test and its result should be entered on the baby's record card.

In a very small proportion of phenylketonurics, early morning specimens of urine are free from phenylpyruvic acid. It is therefore best to test urine passed within a few hours of a meal.

SUMMARY

Phenylketonuria causes grave intellectual and neurological deterioration unless treated with a diet low in phenylalanine. To be fully effective this treatment must be started within a few weeks of birth, before any clinical signs appear. The urine of all babies should be tested for phenylpyruvic acid so that, in affected infants, treatment can be started early enough. The test is very simple, and can be carried out by a health visitor, midwife or nurse, using a wet napkin. Positive cases should have the diagnosis confirmed by a laboratory with the least possible delay before starting treatment. It is best to test

twice, at two weeks and at six weeks of age. If only one test is possible, three to four weeks is the best compromise.

In addition to the newborn, all mentally retarded, psychologically disturbed, or epileptic children should have their urine tested for phenylpyruvic acid. The information obtained is necessary for genetic counselling, and older phenylketonuric children are often improved, to some extent, by a low phenylalanine diet.

RESUMÉ

Recherche de la phénylcétonurie

La phénylcétonurie entraîne de graves lésions de l'intellect et du système nerveux si l'on ne prescrit pas un régime pauvre en phénylalanine. Ce traitement ne saurait être totalement efficace que s'il est entrepris dans les premières semaines qui suivent la naissance, avant l'apparition de tout signe clinique. On devrait rechercher l'acide phénylpyruvique dans l'urine de tous les nourrissons afin que ceux qui sont atteints puissent être traités à temps. L'analyse est très simple et peut être faite par une assistante visiteuse, une sage femme ou une infirmière à l'aide d'une couche trempée. Avant de commencer le traitement, les cas positifs devraient être confirmés, dans les plus brefs délais, par un laboratoire. Il est préférable de faire deux analyses, aux âges de 2 et 6 semaines. Si l'on ne peut faire qu'une seule analyse, le meilleur parti à prendre est de la faire à 3 ou 4 semaines.

Outre le nouveau-né, on devrait faire analyser les urines de tous les enfants débiles mentaux, atteints de troubles psychologiques ou épileptiques. Les résultats servent aux consultations d'ordre génétique et un régime pauvre en phénylalanine amène souvent, dans une certaine mesure, une amélioration chez les enfants phénylcétonuriques.

ZUSAMMENFASSUNG

Proben für Phenylketonurie

Phenylketonurie ruft schwere intellektuelle und neurologische Störungen hervor, wenn sie nicht durch phenylalaninarme Diät behandelt wird. Um einen völligen Erfolg zu erziehlen, muss diese Behandlung in den ersten Wochen nach der Geburt anfangen, bevor irgendein klinisches Zeichen erscheint. Der Harn aller Säuglinge müsste auf Brenztraubsäuregeprüft werden, so dass man bei befallenen Kindern, die Behandlung zu rechter Zeit anfangen kann. Die Probe ist sehr einfach und kann von einer Fürsorgerin, Hebamme oder Krankenpflegerin mit einer nassen Windel durchgeführt werden. Positive Fälle müssten so schnell wie möglich vor Beginn der Behandlung von einem Laboratorium bestätigt werden. Es ist besser zwei Prüfungen, im Alter von 2 und 6 Wochen, vorzunehmen. Wenn man nur eine Prüfung machen kann, ist es zweckmässig, sie im Alter von 3 oder 4 Wochen zu machen.

Ausser bei Neugeborenen müsste auch bei allen geistig zurückgebliebenen, psychologisch gestörten oder epileptischen Kindern eine Harnuntersuchung durchgeführt werden. Die Ergebnisse sind für genetische Beratung nützlich und ältere Kinder mit Phenylketonurie verbessern sich oft, bis zu einem gewissen Grade, durch eine phenylalaninarme Diät.

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Music Therapy and the Cerebral Palsied Child

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THE therapeutic value of music lies in the fact that it communicates through the senses, and it affects the mind, the body and the emotions at different levels of consciousness, irrespective of age, intelligence or education. Music seems likely to be successful with the cerebral palsied child because it can reach him physically, mentally and emotionally. It can be a useful adjunct to physiotherapy, speech therapy and psychotherapy.

Research

Little scientific research has been undertaken so far into the effect of music on the cerebral palsied child. Lack of funds and difficulty in forming a team of specialists may account for this since both musical and medical knowledge are required. It is hoped that this paper may evoke interest in this new subject and help towards the planning of a research project.

There has been very little study of the relationship between medical diagnosis of the different types of cerebral palsy and the reactions of the patients to music. Research in this field could be valuable, as can be judged from an experiment conducted by Dr. Erwin H. Schneider (1956) at the University of Tennessee.

With each of the two main groups of cerebral palsied children—spastics and athetoids—he used music of different kinds, which he called 'physically stimulative' and 'physically sedative'. The distinction between these two types has been defined by Dr. G. Thayer Gaston (1951) of Kansas University:

> 'Stimulative music is the type that enhances physical energy, induces bodily action, stimulates the striped muscles, the emotions and the subcortical reaction in man. It is based on such musical elements as strong rhythms, volume, cacophony and detached notes.
>
> 'Sedative music is usually of a sustained, melodic nature in which strong rhythmic and percussive elements are lacking. This results in sedation, and responses of an intellectual and contemplative nature rather than physical.'

Each group of children was asked to perform two psycho-motor tasks in two different situations:

(1) Colouring geometrical patterns;

(2) Putting pegs in a peg board;

during which either sedative or stimulative music was played on a record. The children were conscious of the music but did not listen to it actively. A few of them became aware of the effect of the music on their work.

The results were briefly as follows:

During sedative music the athetoid children showed improved control and less distractibility. The reverse happened with the spastic children, whose performance seemed to be impaired by the effect of sedative music.

During stimulative music the athetoids' motor control became poorer. They exhibited more generalised motor activity and excitability. On the other hand, the performance of spastic children improved and they showed greater motor control and attention.

This experiment seems to confirm the assumption that there is a correlation between the type of cerebral palsy and the way music affects the child. We must therefore exert great caution when we use music with mixed groups of cerebral palsied children if we wish to motivate certain tasks or activities, or to induce certain moods. In music therapy it would be best to group the children according to medical diagnosis.

Further research of this kind could perhaps elicit useful diagnostic information. The reactions of specially selected subjects to different kinds of music may throw additional light on motor control tests.

Active Listening and the Effect on the Child of Various Sorts of Music

Background music exerts an indirect effect on the child. When he is making music, the effect is more direct. Complete direct impact will take place when the child listens actively to music, especially if it is a live performance in an intimate setting. Then, he may feel deeply satisfied.

I think from experience that the best music to play to a cerebral palsied child is that which holds attention and produces enjoyment without tension. Music is almost always emotional and emotional tension may induce physical tension. Provision of the right emotional climate depends much on the kind of sound produced by the instrument, irrespective of the piece or the composer. The sound of a wind instrument such as the flute, the oboe or the recorder gives a feeling of active, continuous but effortless movement which provokes attention and gives pleasure without arousing tension. The tone of the piano or even of a string instrument is not always ideal for this purpose and has to be graded carefully. I have noticed that the tone of a muted string instrument is soothing and emotionally effective with cerebral palsied children. They seem to find relief and satisfaction in an emotion which can touch them without provoking the tension that so often accompanies their other emotional experiences.

On the whole, a piece of music for solo wind or muted string instrument would be classified as 'sedative' unless it was played very fast or contained sudden changes of pitch. High speed and unexpected contrasts in the music produce a physical and emotional tension in the listener, which is different from the physical response to physically stimulative music. This tension may be more than the child can take, and may then evoke undesirable effects, such as restlessness, excitability or uncontrolled behaviour.

Physiotherapy

A small but keen number of physiotherapists are using music in the treatment of cerebral palsied children, but their musical knowledge is too limited to carry them very far. They have observed that music contains a basic rhythm that can provoke a spontaneous physical response. The basic rhythm of music is called 'pulse'. It is an accentuation recurring at regular intervals, which has sometimes been compared with the regular rhythm of the body. Almost any child can respond to musical rhythm, and even a cerebral palsied child may do so within his physical limitations. I have observed many of these responses, and although they are sometimes very weak, one can work on and develop them.

The most elementary response is beating time to the music, which can be done with the head, with a finger, with any part of the body, in any position, sitting, standing or lying down. Many tunes consist of a simple regular pulse or accent in the bass, and a melody. The blending of melody and rhythm gives a feeling of regular, continu-

ous movement. As examples of suitable and unsuitable music for the purpose, a traditional minuet would provoke movement without increasing excitability; a reel would be too strong a stimulant. The music should first fit the type of response required from the child and the condition of the child. On this basis, the physiotherapist could direct the child's response towards the specific exercises to be done with music in the background.

Music possesses rhythm and duration which can be linked with motor control in time and space. The regular recurrence of the accent and the continuity of the melody may help the child to co-ordinate his movements, to make a more sustained effort and to stretch perhaps further than without music. This fact has already been observed (Mee 1959) in the physical rehabilitation of adult patients at the Camden Road Medical Rehabilitation Centre, London.

The physiotherapist should assist the child when he is making music himself. For instance, when he plays in a little percussion band, his movements should be guided when he is prompted to use his hands, a limb or part of a limb, to hold an instrument or to move with the music. During such activities music should be chosen which will produce the desired effect, and here the musician's advice is needed.

Results are obviously better when musician and physiotherapist co-ordinate their efforts. At the Occupation Centre of Altrincham Spastics Society, some musicians and the physiotherapist are now working together. They try to use music of the right type as a means of relaxation before and during treatment, and as a means to motivate and co-ordinate movements during physical exercises and games. They also use music as a means of a communication with the emotionally disturbed and severely retarded children.

Speech Therapy

Speech therapy is often part of the treatment of the cerebral palsied child. Excellent results with specially chosen songs and ditties have been obtained at the Speech and Hearing Clinic of the North Western University of Chicago (Westlake 1951). The British speech therapist, Miss J. M. Turpin (1959), believes that speech defect often comes from an arhythmic disposition, and uses music to develop a basic sense of rhythm and co-ordination. At the Speech Clinic of the University Hospital in Utrecht, music is also used with cerebral palsied children suffering from a lack of physical rhythm. The performance of seriously ataxic children moving their bells or drum sticks rhythmically in the so-called orchestra has surprised medical observers. (Van der Hurk, pers. comm.).

Mental Development—Mental Retardation

For successful treatment of the cerebral palsied child we do well to take the whole being into consideration; this may explain why music, affecting as it does body, mind and emotions, is often effective in remedial treatment and education. Moreover, music can be perceived and enjoyed at a very low level of intelligence, since it is non-verbal communication and does not need a high mental process to be apprehended. Palmer (1952) and Weigl (1956) give instances of severely brain-injured people who were able to respond to music in an almost normal way. This may be the only way in which we can reach and communicate with a child suffering from multiple handicaps. In numerous experiments on the effect of a live performance of music on the mentally retarded, from the dull child to the imbecile, I have found that the emotional impact of music can act as a definite stimulus to the mind and can then help towards the development of sense perception and mental activity.

These experiments (Alvin 1959), conducted in various schools or occupation centres, have shown that children attending regular music sessions developed a gradual improvement in awareness, span of attention, memory and verbalisation.

Social Integration

Music is an admirable means of social integration with groups of children, either playing or listening. This fact has long been noticed by educationists, especially in the case of handicapped children whose life is bound to be impaired or limited. The following description (Alvin 1958) of such work carried out in the Cerebral Palsy Unit at Queen Mary's Hospital for Children, Carshalton, will give an idea of what can be done with a very mixed group of young children.

'The teacher of cerebral palsied children faces an arduous task. The nature of the illness puts severe limitations on the physical and mental development of the patient. This makes normal contacts and communication difficult. But music reaches these children and can give many of them various emotional and mental experiences that they could not get in any other way.

'In this ward, I found 14 children from 5 to 11 years old, ready for their music lesson, sitting in a semi-circle round the piano. Several of them had no speech, most of them had no control over their movements, some could not use their hands or their lower limbs. But there was in the group a feeling of happy expectation for the music lesson.

'The teacher gives the children small percussion instruments, and tries to provoke physical response to musical rhythm. An assistant helps the children unable to hold or to move their instrument. Some of the children have bells tied to their arms of feet. They all try to follow the tune played on the piano by the teacher. She attempts to give them a feeling for melody and rhythm. I noticed that the physical response to rhythm was good, and that the children enjoyed the proceedings immensely. Without being prompted, children helped one another to pick up or to get hold of an instrument. The emotional and social benefit of this group activity was evident. There was also some singing although many of the children could not emit a musical sound.

'The teacher tells them simple stories about music, for the benefit of the more intelligent children. Some of them tried to repeat to me with eagerness a few facts they remembered about Beethoven's life. The teacher uses the piano during the lesson, and also puts on some gramophone records. As I expected, only a few of the children were able to listen attentively, but the music made some general effect.

'Music is a good medium to give these deprived children pleasure, happiness, stimulus or the feeling of a presence. This music lesson in the ward was an active experience in which each of them could participate. Once more I was convinced of the power of music to reach the deprived mind or body, and I felt how much could be learnt by a serious study of the use of music with cerebral palsied children.'

Emotional Adjustment

Music making or listening may be a substitute for impossible tasks to a cerebral palsied child who has to face and to accept his disability. If the disability affects the entire child it may help him to go even further and overcome this. In music he may find a field of normal activity. An adult cripple told me once that, after a bad accident to his hand, occupational therapy had helped him, but could not make him forget his lost fingers. Only when he started playing music was he able to overcome his disability and to feel normal again.

When music is well adapted to the

child's limited ability, he can find in it a way of expressing himself as a normal child. The emotional impulse may even make him achieve more than was thought possible. Playing an instrument in a band,

Drawing by a cerebral palsied child, inspired by music, and expressing movement.

however elementary, gives a sense of achievement and fulfils the basic need for self-expression. Admirable results have been achieved at the Wilson Stuart School for Physically Handicapped Children in Birmingham by Mr. Geoffrey Smales who has adapted the musical techniques to the physical condition and to the mental and emotional needs of a number of spastic children.

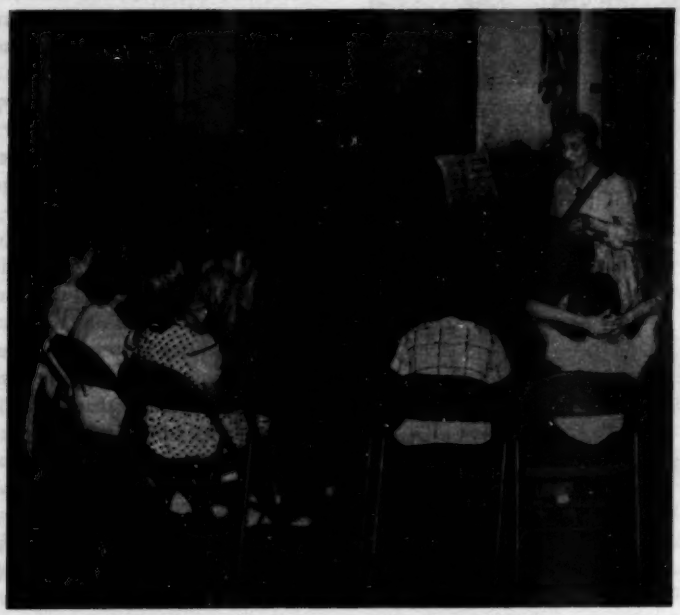
At a higher intellectual level, the study of music can feed and develop the mind and the emotions of the intelligent cerebral palsied child, in spite of his own restricted world, with something that is normally available to any child. He can enjoy music lessons, gramophone records, radio or television concerts. He may even experience through listening to music an emotion of which he is sadly deprived; the emotion of physical movement through time and space, as for instance running, dancing or flying. I have seen pictures made by cerebral palsied children which were inspired by music expressing the emotion of movement. These pictures are spontaneous impressions of emotional release and gratification. Some drawings made by children at Queen Mary's Hospital after a concert are very significant in this respect.

Remedial Education

The learning capacity of the handicapped child is often impaired. Educationists are trying to devise remedial methods to help the child whose intellectual difficulties due to this handicap prevent him from absorbing knowledge in the normal way. These methods may be linked with the therapeutic measures which can develop the child's learning ability. For instance, speech therapy may improve the verbal capacity necessary for school work, physiotherapy may enable him to use his fingers for writing, psychotherapy may help him towards the mental stability necessary for intellectual development. This remedial education is undertaken by teachers specially trained to understand the child's handicaps and the medical treatment he is receiving. Music is now used extensively in the remedial education of the cerebral palsied child. In special schools or occupation centres, it often plays an important part in the time table, especially if a good specialist is available, who understands the children's needs and limitations.

A few reports of valuable work have been published in this country. Lubran (1958) described the work done at Irton Hall School (National Spastics Society)

by the Principal and a team of teachers, which shows the beneficial effect of music throughout the school. This music is one of the few school activities which can be used in many similar ways with normal or handicapped children. It can be fitted in the curriculum to suit the need, the mood or the occasion. At Irton Hall, it is used during Assembly, at rest time as an inducement to relaxation, as a help to physiotherapy, as a social and group activity, and as compensation for immobility. The children listen to or play music of different kinds and for different purposes as do normal children. The Principal, Mr. A. Lubran's, experience and observations confirm other reports made here and abroad on the beneficial influence of music on cerebral palsied children. He has noticed (Lubran 1960) that the skilful use of music can provide emotional release, induce shared activity, encourage movement and assist mental maturation. This educational work with music is based on the medical researches of G. C. Gordon, F.R.C.S.



The author playing to a group of handicapped children.

Mental Health

A cerebral palsied child, unable to lead a normal life constantly faces problems of social, physical and emotional adjustment. He may become frustrated, maladjusted, depressed, pessimistic or aggressive. His mental balance may be badly affected.

In music he is able to find help towards mental health, an emotional outlet, social activity within his ability and a sense of achievement on which he can build self-confidence. Music can also offer him a beneficial atmosphere of relaxation and calm.

Conclusion

A little scientific research has been carried out on the effect of stimulative and

sedative music on the two main types of cerebral palsied children. The door is now open on a field of observations that could be valuable in treating the disorder. Music is already recognised as a useful adjunct to physiotherapy. In the remedial education of the cerebral palsied child, music has made a positive contribution. It now remains for musicians to join the team of all concerned with the treatment, the education and the mental health of the cerebral palsied child.

SUMMARY

Music can help the cerebral palsied child confront, understand and overcome his problems of physical adaptation, both emotional and social. With skilful guidance, he can find an emotional freedom in music, together with cultural and intellectual experience which he could not find elsewhere. Music can help him find and keep his mental equilibrium, and can thus make a real contribution to his education. Music therapy is therefore being used increasingly in special schools and centres both in England and abroad.

RÉSUMÉ

Thérapie par la musique pour les enfants infirmes moteurs cérébraux

La musique est une expérience affective qui peut aider l'enfant infirme moteur cérébral à affronter, voire, à surmonter ses problèmes d'adaptation physique, affective ou sociale. Adroitement guidé, il peut trouver dans la musique une libération émotionnelle, une activité sociale, des expériences culturelles et intellectuelles qu'il ne pourrait trouver d'aucune autre façon. Bref, la musique peut l'aider à garder ou à trouver son équilibre mental.

De par aspects sociaux, affectifs et culturels, la musique constitue un apport positif à l'éducation de l'enfant infirme moteur cérébral. On lui fait largement appel dans les écoles et les centres spécialisés, aussi bien en Grande-Bretagne que dans d'autres pays.

ZUSAMMENFASSUNG

Therapie durch Musik für Kinder mit Zerebrallähmung

Musik ist ein affektives Erlebnis, das dem Kinde mit Zerebrallähmung helfen kann, den Problemen, welche Ihm seine physische, emotionale und soziale Anpassung stellt, die Stirn zu bieten oder sie sogar zu überwinden. Unter geschickter Leitung kann es in der Musik Befreiung der Gefühle, soziale Tätigkeit, kulturelle und intellektuelle Erfahrungen finden, die es auf keine andere Weise gefunden hätte. Kurz, Musik kann ihm helfen, geistiges Gleichgewicht zu behalten oder zu finden.

Durch ihre sozialen, affektiven und kulturellen Eigentümlichkeiten bringt Musik der Erziehung des Kindes mit Zerebrallähmung einen positiven Beitrag. Man gebraucht sie häufig in speziellen Schulen und Anstalten in Grossbritannien und anderen Ländern.

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A Note on the Postnatal Development of the Human Cerebral Cortex

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WHEN Hill, in 1955, reviewed the electroencephalographic evidence of maturation he and other speakers in the discussion that followed deplored the absence of reliable morphological data which could be correlated with the EEG findings. The name of J. Roy Conel was not mentioned on that occasion although, since 1939, this author had made a major attempt at closing the gap. So far, he has published 6 monographs which deal respectively with the cortex of the newborn (1939), of the one month old (1941), 3 months old (1947), 6 months old (1951), 15 months old (1954) and the 24 months old infant (1959).

This work was carried out while Conel held the appointments of Professor of Anatomy at Boston (from the 4th volume as Emeritus) and of Research Associate in Pathology at paediatric hospitals at Harvard and Boston—an ideal combination for undertaking this research. Altogether 39 brains—fairly evenly distributed over the different age groups—have been investigated. They were chosen for the absence of appreciable cerebral pathology and were fixed *in situ* by intracarotid injection of 10 per cent neutral formalin within a few hours of death. Blocks were cut from numerous cortical areas (Conel follows von Economo's chart and symbols) and sections stained with Cresyl Violet (for cell bodies), Weigert (myelin) and Cajal and Golgi-Cox for nerve cells and their processes. The author is fully aware of the pitfalls of the two last-mentioned methods, but is confident that representative information can be obtained by constant comparison between the two. Numerous illustrations of the findings in the chief architectonic areas are provided: Golgi-Cox is illustrated both by the original microphotographs and by drawings.

In tracing progress from stage to stage Conel relies mainly on 9 criteria which include the weight and length of brain, the width of cortex and of each layer, the number and size of nerve cells, the stage of development of Nissl bodies and neurofibrils, calibre and length of axons and dendrites, degree of myelination and number of pedunculated bulbs or thorns. Progress is not equal in all brains of the same age period, and the not inconsiderable variations are frankly stated; conclusions are drawn from tendencies in the majority of cases. There is a good deal of repetition in all volumes which has the advantage of making each more or less self-contained.

The Newborn

The most notable feature in the *newborn* is that the cytoarchitectonic lamination of the adult brain is clearly established. Large and extra-large cells are present in layer V, including Betz giant cells in the precentral gyrus (FA). Horizontally placed cell bodies are found not only in layer I (where they are known as Cajal-Retzius cells), but in all layers, and they are most

numerous in the calcarine cortex (OC) and uncus. Silver methods reveal a sparse mesh of fibres in every area, densest in layers IV, V and VI. Nissl bodies and neurofibrils are generally missing in the cortex, although both are fully developed in brain stem and spinal cord. Only in some Betz cells does early agglutination foreshadow the formation of Nissl bodies, and some rows of granules are the precursors of neurofibrils. Evidence of myelin in the cerebral cortex is found only around a few vertical fibres of FA. According to all criteria FAγ is clearly leading in differentiation; the area for upper trunk, scapula and humerus is more advanced than those for hand, head and lower extremity. PB and PC (post-central gyrus) follow closely and then, in decreasing order OC, LD and LE (limbic region), HD and HE (hippocampus) and TC (auditory cortex). These areas (almost all motor or sensory cortical stations) seem to be *foci of development* for their respective lobes, well in advance of the surrounding (association) cortex. The anterior frontal region is stated to be the least developed in the whole isocortex.

At 1 month

At the age of *one month* the brain is less gelatinous and its weight and length have slightly increased. The width of the cortex has also increased, particularly in layer V, where the size of the large nerve cells is greater. The number of nerve cells is slightly less, however. Definite flakes (as precursors of Nissl bodies) and rows of granules (primitive neurofibrils) are observed in the giant cells of FAγ which also in length and calibre of axons and dendrites, myelination and quantity of pedunculated bulbs are ahead of PB and PC, OC, TC and HD, in that order. It is interesting that feeble EEG waves, elicited from the anterior central region, begin to appear at the age of one month. Conel concludes, mainly from the work of Gesell *et al.* (1940) and of McGraw (1943) that as a whole, however, infantile behaviour is still regulated by subcortical centres.

At 3 months

Development continues in much the same manner in the brain of the 3 *month old* infant: i.e. increase in weight and length, in width of cortex (particularly in layer I), size of nerve cells, which are again slightly reduced in numbers, increase of calibre and length of fibres, myelin and pedunculated bulbs. The apical dendrites and other vertical fibres ascend to a greater height than in the one month old. Definitive Nissl bodies begin to appear in Betz cells. Horizontal cells and their processes are still present in layer I, but they now show conspicuous signs of degeneration: the tangential fibres of later stages cannot be related to these cells. All these features are most noticeable in FAγ (in the areas for upper extremities more than in those for head and lower extremities) followed closely by PB and, to an almost equal degree OC, but less by TC. In the frontal region anterior to FA some advance is noticeable in the posterior end of the middle frontal convolution (the eye field), but orbital regions and the frontal pole (FE and FF) lag behind. The anatomical findings agree, in general, with what is known of the behaviour of the 3-month old infant when he is at the end of the inhibitory phase of the grasp reflex, begins to repress Moro's reflex, reacts to pin-pricks, and has some visual and auditory perception. Function at this period, however, would be more or less confined to the inner layers of the cortex.

At 6 months

Significant features in the cortex of the 6-*months old* infant are: Ending of the apical dendrites of layer V extra-large nerve cells in the tangential layer; increase

of tangential fibres and of vertical and horizontal fibres in layers IV, V and VI. Increased differentiation of the inner part of III may be a pointer to greater power of association.

At 15 months

The next volume (No. 5) deals with the cortex of the 15-*months old*. Layers II–IV show the greatest consistency in growth throughout the isocortex. The size of the nerve cells continues to increase, while their number gradually decreases. Nissl bodies are found now in many cells, though most markedly in the large cells of V. Rows of granules or matured neurofibrils are commonly seen in nerve cells in layers V and III. No lipofuscin pigment is encountered anywhere. Dendrites and axons of all neurones continue to grow, more apical dendrites ending in layer I in which the tangential fibres have also increased. More fibres stain with Golgi-Cox than with Weigert, myelination lagging slightly behind. Pedunculated bulbs have also increased in numbers—especially in the inner layers. No direct contacts of axons with cell bodies, axons or dendrites of other neurones are observed anywhere. Transmission of impulses does not appear to depend upon cell-to-cell contacts; the pedunculated bulbs merely facilitate reception. In this respect Conel's interpretation resembles Bok's (1959) vacuolar hypothesis of transmission. FAγ is still in the forefront of development. Anteriorly, development gradually decreases, being least advanced in the orbital region. PB and OC follow FA closely, while TC lags behind. This is in agreement with the greater physiological importance of vision than of hearing at this stage. OC is more advanced in its anterior parts than at the pole where macular vision is represented.

At 24 months

In the 24-*months old* infant advance is recorded particularly in the outer portions of layer III. Although motor and sensory centres lead, differentiation has markedly spread to anterior parts of the frontal lobe and to the parieto-temporo-occipital 'associational' areas. In some brains, a difference now appears between the right and left hand areas in FAγ: At the age of 24 months the infant is on the verge of handedness. He walks and runs, though still awkwardly, uses short words and sentences, is under way to control such acts as sucking, crying, tantrums, excretion, but is yet unable to oppose the thumb. All behaviour, including morals, is ultimately based on the use of muscles (for the benefit or harm to himself or others), and one of the functions of the anterior frontal lobe may be to control the activities of the motor cortex.

Illustrations

The *illustrations*, both macroscopic and microscopic, are of the highest quality and well chosen throughout. They illustrate particularly well the growth in length and calibre and the progressive myelination of the endogenous and exogenous nerve fibres. One should, for example, compare Figs. 27 and 28 in Vol. 1 with Figs. 15 and 16 in Vol. 3 and Figs. 15 and 16 in Vol. 6. They demonstrate, in Golgi-Cox stained sections and drawings, the remarkable growth of the mesh of fibres in the hand area of FAγ; particularly in the inner layers. With the permission of author and publishers, the drawings (Fig. 28 and Figs. 16) have been reproduced in Figs. 1–3 of this note in order to illustrate this impressive difference. The progressive myelination is well shown in microphotographs of Weigert-stained sections in Vol. 2, 3 and 5. In Vol. 2, the differences in each subregion of FAγ are clearly seen in Figs. 211–214 and can be compared with an altogether less advanced stage in, for example, the superior frontal convolution

(Fig. 215). Likewise there is in Vol. 3 a clear difference of myelination between FAγ and FB in Fig. 202, and between FB and anterior frontal and inferior parietal convolutions (Figs. 209–214). Figs. 202–233 of Vol. 5 show an incomparably more advanced stage of myelination throughout the isocortex. The difference in Cresyl-Violet-stained sections are not nearly so dramatic, since the final lamination is already reached at the stage of the newborn. Here micro-counting and micro-measuring are required to assess significant differences.

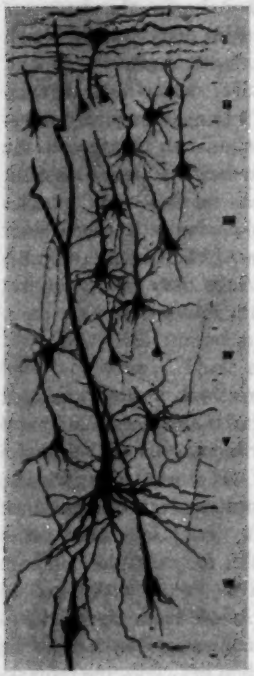

Fig. 1

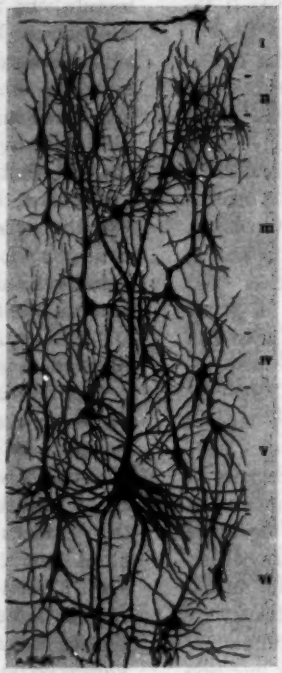

Fig. 2

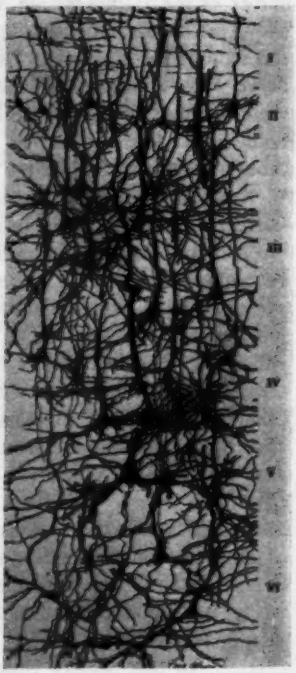

Fig. 3

Drawings from Golgi-Cox stained sections (reproduced from 'The Postnatal Development of the Human Cerebral Cortex'). They demonstrate the progressive development of axons and dendrites in the hand area of the frecentral cortex at birth (Fig. 1), at 3 months (Fig. 2) and at 24 months (Fig. 3).

Conclusions

Professor Conel's work has been summarised in some detail in order to convey an idea of the almost epic quality and dimension of its conception as well as of the painstaking execution and lucid critical interpretation. Although he stands—admittedly—in respect of many findings on the shoulders of Flechsig (1920), Cajal (1909), Lorente de Nó (1943), Langworthy (1933) and many others, he has for the first time singularly succeeded in moulding the older with his numerous new observations into an organic whole. In doing so, he has earned the gratitude of all (anatomists, physiologists, psychologists and paediatric clinicians) who are interested in the problem of maturation.

Some may argue with Conel's views on the olfactory function of the hippocampus, his emphasis on the C.S.F.-brain barrier in comparison with the blood-brain barrier, his views on the function of the frontal and limbic lobes etc. However, these are trifles compared with the magnitude of his achievement, and he disarms potential critics by his modest statement that the 'only reason for presenting them' (his ideas) 'is the hope that some of them may serve as leads for other investigators who by supporting or refuting them will be contributing to our knowledge of the Nervous System.'

In using von Economo's symbols, Conel may have laid himself open to criticisms which have recently been levelled against architectonics, even the moderate ones of Brodmann and von Economo (Bailey and Bonin 1951, Le Gros Clark 1952, Sholl 1956). However, much of Conel's investigation is concerned with areas (the cortical motor and sensory stations), whose architectural definition has never been in doubt, and where he goes beyond these he makes no claim for exact boundaries. In fact, he uses the symbols more or less as convenient landmarks, and as such they will most likely continue to be useful to anatomists, physiologists and neurosurgeons, whatever may be their shortcomings as true maps of the cortex. Conel's concept of cortical function could not be further removed from the 'organologies' of Brodmann (1909) and Vogt (1943). To him, as to Lorente de Nó, the laminar architecture on which most charts are based is not as important as are the vertical striations formed by the apical dendrites and by vertical (endogenous and exogenous) axons. These are the same everywhere in isocortex and allocortex*, suggesting a similar principle of function: i.e. the transmission of impulses without regard to what may initiate the impulses. Any specificity of function of cortical neurons depends, in his view, upon their connections.

The concluding pages of the 6th volume, with their emphasis on fundamental problems of structure and function, seem to portend that this may be the last volume of the series from the pen of Professor Conel. Whether this impression is true or not, it is hoped that the work will be continued at least to cover the first decade of life. Now that plans are at hand for the formation of an International Institute of Brain Research, the postnatal maturation of the brain—not only from the anatomical point of view—may be an admirable project to be tackled there. A work of such dimensions may eventually exceed the capacity of any individual worker; and a combined effort by a number of experienced workers may well neutralise those vestiges of subjective interpretation against which even the most objective single investigator can hardly hope to be immune.

Acknowledgements: I wish to record my gratitude to Prof. J. R. Conel and to Harvard University Press for their kind permission to reproduce the three illustrations.

* Isocortex = fully developed six-layered cortex.
Allocortex = the older more primitive cortex (including olfactory cortex).

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SPECIAL ARTICLES AND REPORTS

Cerebral Palsy at Saint Justine Hospital, Montreal

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THE Cerebral Palsy Clinic at Saint Justine Hospital was organised in 1951, and, after a rather slow beginning, the case load increased so rapidly that in the last four years we have seen over 700 new cases. This unexpected number of cases has led us to revise upwards the classically accepted incidence of the disease from 7:100,000, which would account for only 400 cases in the whole Province of Quebec, to 1:1,000, which thus projects between 5,000 and 6,000 cases. So far, over 2,500 cases have been registered at the Cerebral Palsy Association.

All the cases seen in the Clinic are referred by family physicians or by paediatricians. The only exception is a relatively small number of cases sent to us from outlying parts of the Province by Public Health nurses or through the Cerebral Palsy Association of Quebec.

This Association has been the initiator of the Cerebral Palsy Clinics in our Province. It is due to their untiring work and ceaseless stimulation that our Clinic and that of the Montreal Children's Hospital were set up. Lately, they have assisted in founding similar clinics in Quebec City, Chicoutimi and Trois-Rivières, which will diminish our case load and enable the patients to be treated nearer home.

Staff and Set-up

Our Clinic is composed of an orthopaedic surgeon, a neurologist, a physiatrist, a physiotherapist, an occupational therapist, a speech therapist, a special educator, a social worker, a secretary and a paediatrician, who also acts as Director of the Clinic. Apart from these regular members, the following are always on call to investigate or treat the patients: a psychologist, an ophthalmologist, an audiologist, a dental surgeon and an orthodontist.

From the beginning, our philosophy of treatment has been that we should help the child to help himself in developing his physical and mental faculties to the utmost. We also believe that this should be done in his home, so far as this can be achieved. We do not believe that there is any value in taking the child out of his familial and social environment to train him in an artificial atmosphere, such as exists in special homes or institutes, because these patients tend to regress when they return home, as they must do sooner or later.

We try to habilitate the child to the limit of his potentialities. If he can be raised to the level of independence, then he is fitted into regular or auxiliary classes of the School Commissions. If he cannot reach this level, we direct him to specialised schools, of which there are too few at present; only if he is ineducable or untrainable do we place him permanently in an institution for the mentally defective.

With this aim, each new case is seen at the Clinic, where he is examined by all the members, who then decide the type of treatment to be used. Unless surgery is contemplated, the child will be hospitalised

only if further investigations are needed or if he lives very far away and in a locality where there are no possibilities of the treatment being carried out. Usually, our cases are treated on an outpatient basis, together with a home programme, which is taught to the parents and which is very important in getting the family interested and active in the child's treatment. For the patients living within 15 miles of the hospital free transport is provided by the ambulances of the Quebec Society for Crippled Children.

Now that Cerebral Palsy Centres are being opened elsewhere in the Province our role is changing, at least for the patients from the localities where Centres exist. We are functioning more and more as a diagnosis and evaluation Centre, setting up the plan of treatment for each case, but then referring him back to his own Centre for continuation of the treatment. Some of these cases are seen again for re-evaluation and sometimes for operative treatment by the orthopaedic surgeon or the ophthalmologist. We have found that most of our cases of squint do not gain any fusion by the correction of their strabismus, but the cosmetic results are worth while. Most of these cases are treated initially by occlusion, surgical procedures being used only when there is insufficient correction or none.

Our treatment services were planned to answer all the needs of the cerebral palsied children in one physical unit. This unit has a direct entrance for the transport ambulances and is also connected by elevators to the five floors of wards for inpatients. The Physiotherapy Department has a gymnasium and a large swimming pool and is staffed by 12 physiotherapists. The Occupational Therapy Department is fully equipped for its work and can handle many patients at the same time. It is concerned mostly with the training of the children in the activities of daily living. It teaches their mothers to carry out the same programme at home and helps them in devising or choosing appliances that are useful in these activities.

The Speech Clinic can evaluate and treat any speech disorder, usually on an outpatient basis. The treatments are given at the time of the patients' visits to the Physiotherapy Department.

We benefit also from the co-operation of the Psychiatry Unit, the Audiology Department and the Ophthalmology Department. We are particularly fortunate in having an Orthopaedic Appliances Department, which has been functioning for almost 30 years and where our cases are supplied with braces as needed.

Conclusions

A cerebral palsied child can and must be treated, if he is to attain his maximum development, which will vary with his intellectual level.

To reach this end, the team approach must be used both in the evaluation of the entire child and in his treatment. It is only through the mutual understanding and close co-operation of the many specialists concerned that these patients can realise their full potential.

Even this is not sufficient and the parents must be brought into the treatment plan, so that, understanding the problem of their child, they can take their place in it. They must also understand that these children should not be overprotected or spoiled but should be constantly stimulated and prodded on to work by themselves, this being the only way in which real and permanent results can be obtained.

The Cerebral Palsy Clinic thus becomes more and more a guide and counsellor for the parents, who carry on most of the daily work of treatment. The Clinic itself and the Hospital are used when a re-evaluation has to be made or in medical or surgical emergencies.

Assessment in Cerebral Palsy

Discussion at a Joint Meeting of the Medical Advisory Committees of the N.S.S. and B.C.W.S., at The Hospital for Sick Children, Great Ormond Street, London

THIS discussion was held on February 25, 1961, with Prof. A. A. Moncrieff in the chair. Dr. J. H. Crosland, Dr. Ronald Mac Keith, and Prof. A. V. Neale represented the National Spastics Society, and Dr. H. M. Cohen, Mr. S. Evans, Dr. Errington Ellis, Dr. I. Holoran, Prof. R. S. Illingworth, Mr. A. Innes, and Mr. G. A. Pollock the British Council for the Welfare of Spastics.

The main speakers were: Dr. K. W. Nicholls Palmer, director of physical medicine, Colchester Group Hospitals; Mr. Alexander Innes, M.B.E., F.R.C.S., orthopaedic surgeon to the Children's Hospital, Birmingham; and Dr. P. Hume Kendall, M.R.C.P., senior registrar in the physical medicine department, Guy's Hospital, London.

General Principles of Assessment

Mr. Nicholls Palmer said the subject of his talk had been chosen for him and had proved more difficult than he had expected. The definition of 'assessment' in the Oxford dictionary was not helpful, and the word 'value' seemed more appropriate in the present context.

Assessment dealt with people and with personalities. One should know what one was assessing and why one was assessing a particular person. One might want to know their present condition or their future potential. One might be assessing them for life in their own home, in an institution, in a sheltered workshop, or in open employment. In any case one had to assess their general capability, and Dr. Nicholls Palmer felt that the order of importance was communication, locomotion, toilet, intelligence, mental outlook, and skills and abilities. He showed slides to demonstrate various ways in which handicapped people could communicate and overcome their handicap in unconventional ways, illustrating that the way in which things were done did not matter so much as the end result.

When a person's brain had been damaged, his brain could not be regarded as normal, but he might still be able to adapt himself to his environment. Someone born with only one arm could usually adapt himself and lead a very full life, and would not be likely to have more psychological problems than unhandicapped people. However, a cerebral palsied person might have more difficulty in adaptation, because the brain itself was involved and there might be changes which were difficult to assess. For instance, if such a person had one useless arm, it might not only be useless, but require concentration to control athetoid movements and might continually call his attention to an abnormal structure. Where only one arm was involved, it was often not accepted by the individual, particularly in childhood, and Dr. Nicholls Palmer was not sure whether one helped by assessing its potential and striving to make use of something that had been rejected.

General appearance influenced the person carrying out the assessment. One should see the patient as a whole and treat him or her as such. It was very difficult to assess the effect of involuntary movement or lack of movement on the person as a

whole. One had to assess not only what one found on examination but also potential development, both of the child under treatment and of the adult who had been neglected. A doubtful case should be given long-term assessment in a hospital or centre. This meant observing him when he was living in a community and studying his reactions to other people. When being tested or assessed, a patient might be quite able to weave or use a lathe, but he might not want to do so, or might be able to concentrate for only a very short while. He might be easily distracted and quite unsuitable for open employment. Simple tests were not sufficient; in most cases there should be a period of trial.

Dr. Nicholls Palmer ended by suggesting that one should have a reason for carrying out the assessment, and should use some form of capability chart (not too complicated or time-consuming), some form of recording mental capability, one's eyes and a lot of common sense.

Difficulties in the Assessment of Treatment

Mr. Alexander Innes said that assessment of progress could be made only by a study of accurate records taken over a period.

A child's progress could be considered from two main standpoints—mental or educational, and physical. Of the two, educational progress was the easier to assess, because the simple filing of specimens of the child's writing and number work would afford a continuous picture of the advances made in response to teaching methods. The recording of physical progress—which, in the cerebral palsied child resolved itself into recording either the acquisition of motor skills or their improvement in performance—was a much more difficult task, because it was necessarily discontinuous, taking place at intervals of perhaps eighteen months. Therefore, if at the time of the recording (on ciné film under controlled conditions) the child did not perform up to his usual level, a very false picture would be obtained and a faulty inference drawn when the filmed series was studied.

The factors which might influence the child's performance and so affect the assessment could be divided into (a) extrinsic, in the home or school, and (b) intrinsic, arising from physiological or pathological processes within the child itself. The home background and influence were very important, particularly to the handicapped child. Thus, a 14-year old girl who had been making steady progress in walking had regressed considerably during the Christmas holidays because of her disturbing home background. This extrinsic factor had seriously affected her progress, and if her motor performance were recorded now, the assessment would be one of complete failure of response to treatment, and indeed regression.

The birth of a normal brother or sister could also significantly affect the performance of a cerebral palsied child. First, there was the fear of rejection, which was especially prevalent in the child with cerebral palsy who was essentially dependent for so much on his parents; and later, when the presence of the new baby had been accepted, there was the factor of competition with the younger, normal child. This latter factor might not always be injurious. Competition might spur the handicapped child to greater efforts and might produce a positive gain in that the elder child might be forced, for the first time, to make a real effort to hold his own place. But, not infrequently, the fact that the baby with his motor skills gradually surpassed the elder, handicapped child, despite the latter's efforts, caused severe frustration, resulting in a failure to concentrate or work hard, which was reflected in his educational and physical progress, and might result in a com-

plete withdrawal from the outside world.

Tacit rejection by either parent might also have adverse effects on the child's response to treatment, and this was particularly the case if rejection by one parent caused the other to smother the child with doting care and so isolate it from the outside world, denying it even the limited experience possible to the handicapped child. Quite often, when the cerebral palsied child was the youngest in the family, it was treated by all the other members as a much-loved pet. All its wants were anticipated, so that any stimulation of ambition in the school or clinic was at once nullified at home.

Another extrinsic factor was that produced by a surgical operation, even if this were designed to improve the performance of motor skills—for instance, a gastrocnemius recession to relieve a serious equinus deformity of the foot and ankle. After a period in hospital and fixation for some time in plaster, motor skills acquired before the operation might be in abeyance for a considerable period, and it might take some months to get back even to the pre-operation level of performance, although by correction of the deformity this level would ultimately be surpassed and further progress achieved. Even a minor operation might have an effect on the child altogether out of proportion to its status as a traumatic event. This disturbance and retrogression in performance was partly a result of the enforced separation from the family but it was partly due to alterations in the mechanics of the spastic limb by operation or fixation. For a period in plaster would have a direct effect on motor performance which was not simply a result of the postoperative stiffness or limitation of movement but was also due to the alteration in sensory patterns arising from the changes in muscle tension and limb posture produced by the operation.

Turning to the intrinsic factors, intercurrent disease would often retard a child's progress, and childish ailments like severe whooping-cough, measles or mumps might cause further degeneration in the central nervous system. The partial asphyxia occurring during paroxysms of whooping-cough might cause deterioration in a brain already damaged by anoxia, and virus deseases might produce complications such as encephalitis, which were serious enough in the normal child but might be disastrous in a case of cerebral palsy.

Any infection which caused a toxaemia, associated with a moderately high pyrexia, might trigger off an epileptiform convulsion, and after such a fit the child's performance level might be low for weeks and he might regain only slowly his motor skills after unremitting therapy. Normal physical growth also might affect response to treatment in more than one way. Simple obesity could be an obstacle to gaining a good walking pattern. The vicious circle of increasing obesity and decreasing physical exercise was extremely difficult to break, especially in the handicapped child. Apart from obesity, the steady gain in body-weight which occurred as the child matured into the adult might prove too much for the relatively weak extensors of hip, knee and ankle, and children who, when young and light in weight, appeared relatively normal on standing or walking, often showed at the age of 17 an increasing flexion deformity of the hip and knee which was impossible to control except with calipers. These were difficult to manage, when out at work, and were consequently discarded, with the result that the young man or girl shuffled about with knees and hips virtually fixed in a position of 90° flexion.

The normal increase in the length of the long bones also produced mechanical problems which were perhaps not always

fully recognised. Children showed periodic variations in the rate of growth of the long bones, peak increases taking place somewhere between 8 and 9 years and 15 and 16 years, such periods of rapid growth alternating with interludes during which the body weight increased but the increase in limb length was slow. During these periods of rapid growth in length, the normal child developed a tendency to a generalised physiological muscular slackness or hypotonicity; it was at these periods that orthopaedic surgeons saw normal children with postural deformities, such as flat foot, kyphosis, postural scoliosis, and so on, these conditions being an expression of the general lack of muscle tone.

In the spastic child, however, the spastic muscle groups did not relax in this way, with the result that there was a very marked tendency at these times for the flexion-deformities of the hip and knee and the equinus deformity of the ankle and foot to increase, simply because of the relative increase in length of the tibia and femur as compared with the non-relaxation of the spastic calf and hamstring muscles. Records made at this time might show increasing deformity despite treatment, and an incorrect assessment would be made unless this aspect was borne in mind.

The onset of puberty could also have a good or bad effect on the child's performance. In some, especially boys, ambition appeared, which was often lacking previously, and this might result in rapid progress in the acquisition of new motor skills, or in the improvement of the performance of the old. In many children, however, the new awareness and appreciation of their disability, as compared with other happier, normal children, might produce extreme frustration and a sense of hopelessness which might have a very serious effect on their attitude and their response to therapy.

Everyone was aware of the importance of early diagnosis in cerebral palsy, yet even the very elect made mistakes, and, from time to time children could be seen in schools and clinics who had a progressive degenerative condition of the central nervous system which had not been recognised as such, the motor deficiency being ascribed to cerebral palsy. Such cases might show an initial response to treatment but records would reflect their gradual deterioration, and, unless the intrinsic factor responsible were known, a faulty assessment of the results of treatment would be made.

These were not all the variables which might materially affect the response of the cerebral palsied child to treatment, but they were factors of which the physiotherapists were only too aware and which should be taken into account when the results of treatment came under review.

The Problem of Relating Assessment to Treatment

Dr. Hume Kendall said that the published work on the treatment of the motor disability associated with cerebral palsy revealed a wealth of impressions and recommendations on procedure which were seldom accompanied by sufficient detail or by facts and figures in a form suitable for statistical analysis. It had been considered impossible to describe the defects that result from cerebral palsy in a way that could be recorded quantitatively. Yet such an objective assessment was essential in order to answer the main problems that confronted those concerned in the establishment of effective therapy regimes. Among the most pressing problems were:

(*a*) What percentage of cerebral palsied patients achieved economical and physical independence? Could such an objective be hastened or its attainment made more likely by therapy?

(*b*) Which were the most effective methods of management; did the various clinical syndromes respond in a different way to various forms of therapy?

To answer these problems, carefully controlled trials were required, and, as a basis of such trials, an effective system of assessment was vital. There seemed little doubt that such a system should have a quantitative basis—i.e., each feature should have a score, the sum total of all aspects of the assessment being recorded as a 'motor quotient'. Such a quantitative approach had its critics, but it was the only way in which true objectivity might be attained without requiring the expression of an 'opinion'. Simplicity and reproducability were essential, and once a satisfactory scheme had been developed it would be of great value both to the supervising physician and to the therapists concerned. Furthermore, patients and parents alike appreciated the attempt to secure an honest picture of progress.

There were several proven systems of assessment in existence. Unfortunately, these had not been found altogether suitable for practice in a Cerebral Palsy Treatment Unit, covering a range of ages from 2 to 18 years. Review of these systems is informative inasmuch as it illustrated some of the practical difficulties in relating the results of assessment to therapy.

A. Johnson, Zuck and Wingate[1] were the first to develop a system of tests applicable to cerebral palsy. These were devised in such a way that a 'motor age' was determined for each patient and this was correlated with the chronological age. In practice, the tests suggested had three main disadvantages:

(i) They were too complex for any but children of the highest grades of physical and mental development to attain;

(ii) They extended only to the age of 5 years;

(iii) They were, on the whole, inadequate to give an accurate reflection of the child's true physical state—for instance, it seemed unlikely that the ability or otherwise of a child to trace its finger around a star-shaped apparatus without breaking an electrical circuit was a valid way of assessing that its motor age was 5 years.

From these tests a numerical assessment of 'motor age' was obtained which the authors claimed to be accurately reproducible.

B. Ingram, Wilters and Speltz[2] devised a scale based on Gesell's Development Schedules and modified to some extent by Johnson's tests; this gave two levels—a motor age and a social age. Again, the tests had a range of 4 weeks to 5 years, which made them of limited value. The same criticisms that applied to the Johnson, Zuck and Wingate tests were valid here. The tests were not sufficiently comprehensive; they were too complex in some respects and they were not applicable to patients over the age of 5. The authors noted that the results had been of great value in analysing and planning forms of therapy and in formation and prognosis.

C. Karlsson, Nauman and Gardestrom[3] used an extremely simple and practical system to divide their patients into five classes. Unfortunately, they did not give details of how this was accomplished, except to mention that 'use was made of the Hartwell motor age test' and that from

[1] Johnson, M. K., Zuck, F. N., Wingate K. 'The motor age test: measurement of motor handicaps in children with neuromuscular disorders such as cerebral palsy.' *J. Bone Jt Surg.* 1951, 33-A, 698.

[2] Ingram, A. J., Withers, E., Speltz, E., *Arch. Phys. Med. Rehabil.* 1959, **40**, 429.

[3] Karlsson, B., Nauman, B., Gardeström, L. 'Results of physical treatment in cerebral palsy.' *Cerebral Palsy Bulletin.* 1960, **2**, 278–285.

these tests the patients were classified into these five classes:

(i) No practical limitation of activity.

(ii) Slight limitation of activity.

(iii) Moderate limitation of activity.

(iv) Great limitation of activity.

(v) No useful physical activity.

The system of dividing patients' abilities into 5 distinct levels had found great application in other fields of rehabilitation —viz., muscle testing in poliomyelitis. The authors in this study, emphasised the essential reproducibility and the validity of the figures obtained for statistical analysis.

D. *Functional Assessment Chart* (Prested Hall). Dr. Kendall mentioned this system on account of its simplicity and apparently accurate reproducibility. Devised by Dr. Nicholls Palmer, it comprised 54 tests of everyday activity and was aimed at producing a quantitative assessment of the ability of the severely disabled adult with cerebral palsy. This system had proved completely reliable in this group of patients and demonstrated quite small degrees of improvement or deterioration in conditions which corresponded closely to the clinical impression. However, the tests were not applicable to young patients.

E. *Cheyne Walk*. This system seemed the best of the various methods of assessment now available. It comprised seven charts relating to different aspects of function and including several everyday tests, speech tests, etc., all of which were briefly condensed on a single 'key' chart. Unfortunately, there was no numerical scoring for this system of assessment,but it was possible to obtain a vague estimate of the child's ability by studying the shade of the graph on the 'key' chart. The absence of numerical scoring and the limited age range were probably the only deficiencies in this system.

In practice there were several ways in which all the present systems of assessment were defective, at least for those concerned with physical rehabilitation. Firstly, no record was made of specific limb and joint functions. For example, the development of contractures, alteration of spasticity, or changes in spontaneous movements could not be demonstrated by the tests of ability on which most of these systems depended. Thus, by developing greater skill and trick movements, it was possible for a child apparently to improve its motor age on testing while simultaneously developing joint contractures. However, in later life the contractures would prove a great disability and the practical everyday management therefore needed to give attention to the contractures. Secondly, children of limited intelligence frequently could not attain any worthwhile score in the ability test, and therefore the record would suggest that they had not improved from physical treatment. However, simply by increasing joint movements, muscle power, etc., it was possible to make patients far more independent, despite the fact that they could not complete complicated ability tests.

All systems of assessment should at least take specific cognisance of the following features: (*a*) upper limb function; (*b*) lower limb function; (*c*) spatial discrimination; (*d*) posture and locomotion; (*e*) ability tests; and (*f*) appliances.

It was essential to consider the functions of the upper and lower limb separately; good function of the upper limb was necessary for economic independence, and was therefore the more important of the two. Lower limb dysfunction would also prove a handicap in quite different ways and therefore required separate recording.

Each of these features should include a record of deformity, measurement of range of joint movement, muscle testing, co-

ordination tests and a record of involuntary movements. Ability to appreciate different forms in space constituted a very critical test of function, the pathways of which were probably not tested by the usual simple skeleto-muscular examination. In France 'l'age gnosique' was viewed very highly as giving distinct information on the patient's progress: Tardieu (personal communication) has spent much time in developing an excellent system of testing this faculty.

Posture and locomotion should obviously be recorded separately, and the ability to perform these functions should score heavily in any motor assessment. Ability tests were also very important but should constitute only part of the overall picture.

All scores should be divided by an 'Appliance Factor', derived by the assessment of the degree of dependence on braces, etc.

Once it had been decided which physical characteristics were to be tested, there remained two points of difficulty—first, the allocation of scores to the various sections; secondly, the determination of the improvement in the patient's ability that results solely from normal growth. The former was gradually being overcome with experience, but it was difficult to see just how one could account for the progress that accompanied normal growth. To some extent this might be accomplished by comparison of a particular faculty with one of the accepted developmental scales. Unfortunately, this is difficult in patients with cerebral palsy, in which the defect appears to change from day to day and from month to month.

Quantitative assessment of the motor disorder of the cerebral palsied was obviously a difficult and often tedious problem and it was only from constant experience that suitable scales might be developed. The more detailed the scheme, the more valuable it would be in the future, but it was better, at the present time, to have some form of objective record, irrespective of detail, than to eschew this on account of difficulties.

General Discussion

Prof. A. A. Moncrieff said that the Medical Advisory Committee of the N.S.S. hoped that this meeting would reach some agreement on the assessment and results of treatment, just as the Little Club had gone some way towards reaching agreement on terminology. Parents were confused by the sectarianism of treatment—each group, such as the Bobaths, Mrs. Collis, Estrid Dane, saying that they were right and the others were wrong. Until there was some means of assessment which was acceptable to these groups and to doctors specialising in cerebral palsy, it was difficult to give a definite opinion on the different methods of treatment.

Dr. K. S. Holt wondered if ciné-film records were thought to be better in monochrome or in colour; workers in Oxford felt that colour film gave much more accurate results. He also wondered how much physical progress was the result of the physical side of treatment and how much was an educational process of the child relearning movements. These two factors had to be separated somewhere. Long-term planning of treatment and assessment was very important and one had to be prepared to cover the whole range of ages. Assessment should correlate all the different fields of progress, since a child might improve in speech and deteriorate in motor movements. One had to measure a number of factors even in one particular field.

Dr. Holt did not like the use of records which produced a numerical score, for these tended to make one think of a

number rather than of the child as an individual. Figures gave one a false sense of accuracy, and the numerical scores were really not accurate.

The 'Motor Quotient' which had been mentioned was not like the 'Intelligence Quotient'. Most of the doctors using this method of assessment scored motor abilities, and for each ability the patient was given a point. Abilities from birth to the present age of the patient were added up to make the score which was quoted as the Motor Quotient. For the Intelligence Quotient the patient was given a battery of tests designed for a certain age level, and this gave a horizontal rather than a longitudinal result.

Everyone would agree on the importance of careful records but record keeping was very time-consuming and could not be delegated to other workers. The methods of assessment were not fully standardised and it was very difficult to obtain standard conditions for the children, who were easily tired.

Mr. A. Innes agreed that one of the difficulties of numerical charting was that one could measure the amplitude of movement very easily, but that did not mean that the function of the limb had improved. One should be recording the function of the limb. He felt that one could not separate physical from educational treatment. It was all an educational process, and if the child was not intelligent enough he would not benefit from physical treatment to the same extent as the more intelligent child. The personality of the physiotherapist was as important as her methods of treatment or her skill.

In order to keep film records of motor function, one had to film under standard conditions and each film should show the same movements, so that films could be compared with each other. It was very important to keep a card index of the films as they were made. He personally felt that colour film was expensive and did not make much difference to the usefulness of film records.

Professor Moncrieff had been surprised to hear Dr. Kendall say that he regarded the children as mechanical problems. The physiotherapist in charge of the Cerebral Palsy Unit at Great Ormond Street had become something of a child psychologist and was very helpful in getting the mothers to accept the situation. Her knowledge of the background of the children helped in her treatment of the children and in the assessment of the results of treatment.

Prof. R. S. Illingworth said that, besides assessing the effects of one's own treatment, one wanted to compare different methods of treatment, the effect of drugs etc., and it was very difficult to arrange controls, because of the number of variables involved—age and sex of patient, type of cerebral palsy, degree of involvement, I.Q. (as far as one could judge), associated handicaps, amount of previous treatment, effect of surgery, duration of treatment, secondary results from surgery, personality of the patient, attitude of parents, epilepsy and the effect of drugs used to treat it, and so on. These variables meant that one could not match one child with another. All these factors have to be borne in mind when assessing the results of treatment and should be fully examined. Computers might eventually be able to take these variables, translated into figures, into consideration. One needed a test of function and of the time the patient took to achieve various movements. One had to determine whether an apparent response to treatment was really maturation of the nervous system.

Information about the effect of treatment was needed in order to balance the results of treatment with the effect of time lost from school. He felt that there should be very good reasons before a child missed school.

Professor Moncrieff mentioned that some Local Authorities arranged for children to receive therapy at their schools, but as these schools were usually for all types of physically-handicapped children the physiotherapists were not necessarily those most competent in dealing with cerebral palsied children.

Mr. Innes said that some people felt that treatment was not important and that the child would improve as he grew older without treatment, but he had seen cerebral palsied children in India who had had no treatment and who had become small twisted 'balls'.

Dr. Kendall said he did not, in fact, regard the patient as a brainless automaton but that the procedure of assessment made it vital to obtain an impersonal, objective attitude. Many physiotherapists gave more than physical therapy. What he wanted to find out was whether physical treatment was of any benefit, or whether treatment by drugs alone, or combined with other treatment, was of help to 'spastics'. He would like to see the results of treatment assessed by the Pulheems system, which was used in the Army and had proved very valuable there. He and his colleagues at Guy's were working on a similar system. Without such a system one had to rely on people's individual opinions.

Mr. Innes pointed out that the facts and figures of assessment relied on the person making the observation.

Mr. G. A. Pollock felt that one might get to the stage when one was satisfied with the progress of a child as shown on charts, when very little progress had been made by the child as an individual. Many children achieved a degree of physical improvement but did nothing with it, while many who were grossly handicapped could do a great deal.

Dr. R. C. Mac Keith agreed that in partially blind children there was very little correlation between the amount of vision and what they could achieve with it. If one read Professor Burn's recent lecture on placebos, one found that one could get 40 per cent improvement from inactive drugs, but that did not prove that there were no active drugs which gave improvement. It was worth while aiming at finding a numerical form of assessment, even if it took a long while to find a satisfactory method.

Dr. Kendall said that everyone at the meeting used physical therapy, yet three of them said that physical therapy was no good and did not want to obtain facts and figures to find out the effects of treatment. Many 'spastics' were receiving no form of treatment. It was important to find out whether physiotherapy helped. If personal therapy was more beneficial, then people should receive personal therapy. Physical therapy might not be useful for some forms of cerebral palsy but one could not withdraw it until its lack of usefulness could be proved. Children should learn the tests before they were recorded, and the recordings should be made by someone known to the children and who was used to the recording technique.

Dr. Nicholls Palmer felt that the Pulheems system would be the answer to the problem of the assessment of treatment. If the recordings were made by as few people as possible the error was not big and the recordings were useful.

Professor Illingworth did not think that anyone had said that physiotherapy was no use. The assessment of results of treatment had to be translated into figures, but people should remember that the figures could not be added up to make the final answer.

Professor Moncrieff said that some people felt that children sometimes derived more from other aspects of their visits to Cerebral Palsy U nits than from the actual physiotherapy they received there.

Dr. Kendall was horrified by the cost of physiotherapy, which was estimated at £115 p.a. per child, with the very high cost of ambulances. Children should go to local hospitals for treatment, is possible.

Dr. P. Holoran felt that if treatment could be given at school the staff could work as a team and get to know more about the children and their needs and abilities.

Dr. H. M. Cohen said that the Ministry of Health had set up a committee, under Dr. Scott, to discuss training centres for children unsuitable for education in school. A quarter of cerebral palsied children were ineducable. Progress depended on the innate intelligence of the children, and he wondered whether one should insist on therapy for the children in these training centres.

Dr. Mac Keith hoped that the mothers of children in schools and attending clinics were taught to do the physiotherapy movements with their children at home.

Professor Moncrieff said that physiotherapists had to be trained to realise that mothers were capable of learning the movements to show their children.

Dr. Ellis felt that a numerical assessment was needed although it might be very time-consuming. The really big problem was the assessment of treatment. He still believed in physical treatment for cerebral palsied children, but they had reduced the frequency of attendances at the Percy Hedley School. Mothers had periods of residence at the school with their children and he felt it was best if the parents and children did not become too dependent on the staff at the Cerebral Palsy Unit.

Dr. Crosland questioned Dr. Kendall's arithmetic and said that physiotherapy at St. Margaret's School cost £40 per child per annum. Children should not receive too many sessions of physiotherapy, because they had so many things to absorb and learn at school and at home. Parents could be taught to give the children a lot of practical therapy and management at home.

Dr. Evans asked how one could assess the result of physical treatment and leave out the other factors that might have contributed to any improvement. The negative test was the one to value. If the records showed deterioration one could accept that deterioration had occurred and try to do something about it. If the records showed improvement one could not tell how it had happened, because it was due to a variety of factors.

Mrs. M. L. J. Abercrombie said that if you had a child or children treated under methods A, B, C, and D, many of the factors would be common to these methods and actual physical treatment would be different and would sort itself out.

Dr. Kendall said that one simply had to use controls in assessing the results of treatment. One had to deprive a group of children of any treatment, assess them and then introduce treatment in the form of drugs, physical treatment, or something else.

Professor Illingworth suggested that it would be very valuable publicity to obtain a film of the untreated cases in India, mentioned by Mr. Innes, to show how children did benefit from the treatment they received.

Maturation

First European Congress of Paedo-Psychiatry, under the auspices of L'Union Européenne des Pédo-Psychiatres, Paris, September 16-20, 1960

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THIS Congress, held at the New Faculty of Medicine in the rue des Saint-Pères, brought together child psychiatrists of many nationalities and converging disciplines, and laid down the lines of future research. This report is concerned with papers given in discussion periods rather than those presented in the plenary session, which have already been published.*

A survey of all the reports shows that the concept of *maturation*, extending from the neurone to psycho-analysis, and the endocrine glands to scholastic achievement, can be approached from many angles.

Biological Aspect

The first group of papers are concerned with maturation as represented by biological and endogenous development. Some features of this are not well defined, notably the part played by the endocrine glands (J. L. Camus and C. Koupernick, France) and others have been re-assessed (J. E. Gruner, France) in histological studies on the details of cell structure and inclusions, in relation to maturation of the central nervous system. This progress in dynamic biology has been made possible by advances in cyto-chemistry and the use of the electron microscope.

A. Schwenk (Germany), speaking on hormonal changes during puberty, showed that puberty is revealed much earlier than it is clinically by an established increase in urinary androgens from the age of 8 years. Mme. C. Dreyfus-Brisac (France) described cerebral electrical activity in infants, including the stillborn 5-month foetus. She recognises three maturation stages, at 8 months of foetal age, at 3 months post-natal age, and at 3 years after birth. The report of T. P. Simson (U.S.S.R.), who died in February 1960, dealt with studies of higher nervous activity and the development of conditioned reflexes during child development.

The last report of the 'biological series' came from J. Scherrer and his collaborators, F. Contamin and R. Verley (France), on a functional and evolutionary aspect—the maturation of electrocortical responses, motor and neuro-vegetative activities in mammals other than Man. The authors used three techniques—that of evoked potentials (following peripheral stimulation), on the one hand, and on the other of registering motor and vegetative activities in the course of the development of mammals. This report seems to confirm Coghill's theory (acquired differentiation of functions); another important fact is that spontaneous electrocortical activity is slower than provoked activity, which tends to prove that external stimulation contributes to acceleration of develop-

* This summary is based on the full reports of the Congress edited by Service de Propagande, Édition, Information, of 14 rue Drout, Paris.

ment. Finally, contrary to accepted opinion, nerve impulses may travel along pathways that are not completely myelinised. It is also noteworthy that there is no proof of a single plan of ontogenesis in mammals, which underlines the dangers of extrapolation between animals and man.

M. Verdun (France) attached great importance to the correlation between anthropometry somatotyping and emotional states. A. Friedemann (Switzerland) has a multi-disciplinary approach to maturation before puberty and the same multi-dimensional care is shown by F. G. Von Stockert (Germany), who places maturation in a historical context. V. Fontès (Lisbon) takes a holistic view of maturation, which includes both physical and mental development.

At the end of this long road we come to psychoanalysis, but since Freud's day it also has taken the same turning in the direction of the other sciences. Actually, as Mme. J. Favez-Boutonnier (France) showed clearly, even among analysts there is a wide gulf between the culturalists who attach major importance to environmental factors and the strict Freudians, for whom the instinctual dynamics predominate.

C. Launay and his collaborators, A. Doumic and J. Trelat (France), studied maturation (essentially emotional) of the child in relation to the family, and E. Miller (London) the scholastic aspects of maturation which are as much affective as cognitive. S. Ahnsjo (Stockholm), approaching the same problem from another angle, described the broad scope of educational facilities in Sweden, which provides preschool classes in preparation for community life, special classes for the handicapped and for the subnormal child and the partially mental defective.

G. Bollea and A. Novelletto (Rome) tried to define the concept of biopsychological immaturity in the medicolegal sense, and they believe that harmony of differing factors is important. Thus some individuals grow rapidly in one direction and so remain immature and anti-social. R. Dellaert (Belgium) criticises the idea of penal responsibility in children and adolescents, but believes that knowledge of what is right and wrong should be emphasised.

J. Van Lennep (Holland), dealing with the professional aspect of maturation, insisted that the factors inducive to professional success cannot be predicted by simple I.Q. testing, since social intelligence ultimately represents full maturation. He has found that those in whom puberty occurred relatively early made social contacts more readily and possessed greater spontaneous adaptability. The advanced girls and boys were more interested in the arts than the professions, whereas the slower developers were attracted to technical work. The advanced group, when questioned before puberty, wanted to be drivers, airmen, naval officers or engineers, whereas the others were more interested in becoming missionaries, theologians or priests, watch-repairers, mechanics, machinists or bicycle repairers. Generally speaking the advanced group are easy to orientate in the professional plane, while this task may be difficult for the retarded group. Besides these two groups there is another group, which after a good start seems to level off at the age of 20, at a point below that expected, and another group whose growth is slow and continuous and whose members do not blossom before the age of 30. In conclusion, Van Lennep says that when there is even partial deficiency of maturation there is little chance of attaining executive positions.

Psychoanalysis

The last day was devoted to a symposium on psychoanalysis, introduced jointly by two leaders of French psycho-

analysis, D. Lagache and P. Male. The object—the study of the adolescent—has been neglected by psychoanalysts because of their somewhat exclusive interest in the early years. These authors distinguish in the history of puberty the end of the period of latency, and the beginning of a pre-conflictual period (13 years in boys and 11 years in girls). Contrary to H. Deutsch, Lagache and Male consider that this pre-conflictual period occurs equally in girls. The following stage is more clearly recognisable, particularly in boys. It is characterised by a changed parental relationship which usually deteriorates, a tendency to narcissism and introversion, and the beginning of amorous relationships, which are identifications rather than real relationships. Puberty ends with a post-conflictual period marked by a strengthening of the ego and greater ability to compromise. Finally, the authors stressed the psychopathological aspects and the difficulties in psychotherapy of verbal communication in certain prolonged puberties.

P. Male again opened the discussion session of psychoanalysis with a brief but valuable estimate of the positions of the adolescent viewed from a historical point of view and in relation to his true social framework. Lack of harmony arises not only from a failure of synchronised gonadic and somatic maturation, but also from refusal to accept the instinctual and genital development in relation to infantile defence mechanisms. Soule, from information obtained from the care of the Assistance Publique (Council), believes that ideas about trauma in childhood should be re-assessed. Some of these traumata are physical and some psychological, but their common denominator is their intensity and duration. He particularly emphasised the problem of rape, which is frequent in certain strata of society. Its effect is much more serious if it is committed by the father or instigated by the mother. He showed at the end of his paper that classical psychotherapy in such patients had to be changed in the majority of cases in favour of active, interventionist, and supporting treatment. In well-developed obsessional neuroses, analytical treatment is necessary.

Some representatives of the psychodynamic school have appeared to draw closer to the Pavlovian doctrine. One of the most eminent representatives of Soviet psychological science, Prof. Myassichtcheff of Leningrad, tried to discourage them by reminding the Congress that Pavlov's aim was to unite physiology and psychology—recognising after this that the reflex technique did not cover the complexity of the human problem. He considers that it is experience engraved on the cortical circuits which forms our personalities, and not the blind instincts which Freud let loose in the world when he opened Pandora's box of the subconscious.

This declaration produced a certain amount of emotion in the symposium which had been hitherto calm, and Prof. Lagache, who was presiding, pointed out that psychoanalysis owed much to Pavlov's genius, and he regretted that it might not be possible to say as much for Pavlovism and the ideas of Freud.

Attention should be drawn to the contribution of Dr. Schmitz, who being somewhat detached in a stomatology service, was able to explain to us that misplaced teeth could be psychogenic in origin and might finally appear as frustrations at the oral level. There was also an excellent contribution from Alice Doumic, who analysed the disturbed relationship between the mother and cerebral palsied, encephalopathic, or psychotic child. M. Anzieux presented a convincing picture of the therapeutic efficacy of analytical psychodrama with adolescents. The same theme was discussed by Widlocher, who

showed that in the adolescent, psychodrama could give rise to either a stream of fantasy material, or alternately to a close adherence to the role with which the subject identifies himself. In contradiction to this, there is the forward-looking child whose behaviour is dominated by anticipating what he will be tomorrow—his place in society as a man, and his sexual relationships as one member of a pair.

Among other papers, Dublineau emphasised the relationship between maturation and laterality. Luccioni and M. Porot, from Algerian experience, demonstrated that a society that is passing through an evolutionary phase often decisively affects maturation. Danon-Boileau and Lab reported the results of prolonged treatment of students, in whom they distinguished two pathological attitudes—the persistence of intellectual dependance, or apparent maturity masking a frailer spirit. Katz spoke on the diverse factors which retard the maturation of children with cerebral palsy.

Kotcherguina studied the higher nervous activity (by the reflex path) in children who had suffered obstetrical trauma, and thus presented a very complete picture of speech disorders; Soukharewa contrasted the two varieties of schizophrenia observed before maturity—one specifically infantile, and the other in the adolescent which resembles the illness in adults; Pevzner defined the particularities of higher nervous activity in imbecility. Two papers were devoted to enuresis. One was by P. Holman (London) and stressed the time of nocturnal urination, most often from four to six hours after the onset of sleep, which probably indicates the return to an infantile spinal pattern. Brajevic and his collaborators (Belgrade) insisted more on eminently practical psychotherapy aimed at evoking the inhibitory process. Altschulova (London) described her conception of the work of the child psychiatrist in dermatology. We must also cite the contribution of Amado, illustrating in depth the work of an observation centre, that of Warren and Stein, who studied 187 adolescents—107 boys and 80 girls—and who insist that there is no parallel between the intensity of behaviour and scholastic results. Lelord (Paris) presented a picture of the study of sound and light conditioning registered on an encephalogram. It is interesting to note that the speed of conditioning and the evolution of electroencephalographic results are essentially the same in a child over three years and an adult; nevertheless the EEG responses last longer in an adult; in children with behaviour disorders the responses are brief as with an unbalanced adult. Lairy and Netchine applied themselves to the study of electroencephalographic maturation curves measured by the central occipital frequency. This frequency varies little in function with the I.Q.; on the other hand, children with a low occipital frequency have in common slowness, inhibition, passivity, phobic tendencies and enuresis.

Mme. Dormoy is interested in the study of maturation at the moment of transition from oral to written language. Ataide-Schneeberger has met 28 cases of dyslexia out of 572 children in primary schools in Lisbon—the percentage being 6 per cent for boys and 3.6 per cent for girls; he found a frequent rate of retarded motor development and behaviour disorders. On the other hand the diagnosis of dyslexia in the family antecedents is not frequent. Finally, Meyer found a new type of frontal encephalopathy characterised by gross mental deficiency associated with a non-systematic dyskinesia; in these children there is a frontal atrophy, predominantly on the right side, without much electroencephalographic evidence.

Finally, we were shown several films. That made by Mme. Sainte-Anne-

Dargassies, co-director of the Research Centre of the Baudelocque Maternity Unit, has already become a classic. It illustrates in a striking way that prematurity in the absence of cerebral lesions, catches up, finally, with the behaviour of the child born at term, and that this happens at nine months from conception. This film also illustrates the techniques of neurological examination which are so specialised at this age, and which we owe to the collaboration of this author with M. André-Thomas. Soulairac and Shentoub presented cinematographic research into two particularly fascinating domains of psychology—self-mutilation in imbecility, and the behaviour of the child in front of a mirror.

Finally, M. Geber showed a film illustrating the development of two groups of coloured children in Uganda. One group lives in a traditional non-Europeanised environment, and their motor development is particularly rapid in the first few months; the other group is raised under European conditions and their motor development is slower and comes closer, finally, to that of European children. On the other hand, from the third year on, not only do they catch up with their more primitive competitors but surpass them.

National Association for Mental Health

About a thousand delegates, including a large number of doctors, attended the annual conference of the National Association for Mental Health, held in London on March 9th and 10th. They heard the Minister of Health, Mr. Enoch Powell, declare that the hospital pattern in the mid-1970's, with 50 per cent or less of the present number of mental beds, would have these for the most part in wards and wings of general hospitals. Few ought to be in great isolated institutions or clumps of institutions. And Mr. Powell made it clear that he meant just that: the elimination of by far the greater part of this country's mental hospitals as they stand today. 'For the great majority of these establishments there is no appropriate future use,' he said, 'and I for my own part will resist any attempt to foist another purpose upon them unless it can be proved to me in each case that such, or almost such, a building would have to be erected in that, or some similar, place to serve the other purpose, if the mental hospital had never existed there. Hospital building is not like pyramid building, the erection of memorials to endure to a remote posterity. The old Roman virtue of *pietas* becomes a vice when it is directed towards a hospital building.' Speakers from the floor, who included Miss Bessie Braddock and medical superintendents, were not too happy about the Minister's policy thus declared.

So far as the provision for the subnormal was concerned, Mr. Powell conceded that the prospects for change must seem far less dramatic—'certainly if we discount, as we prudently ought, the chance of some medical "break through" on this front'. Far from contemplating the certainty of a heavy run-down in numbers, we had to reckon with the increase which flowed automatically from the increased expectation of life of the subnormal and their improved prospects of surviving infancy. 'I have the impression that we know much less than we ought to about the prevalence, variety and management of subnormality, but, handicapped though we may be by these deficiencies, the hospital plan has here also to be founded on assessment of need which leans as far as possible in the direction of the trends that we wish to favour.'

Mr Powell said a hospital plan made no sense unless the medical profession outside the hospital service could be supported by a whole new development of local authority services for the old, the sick and the mentally ill and mentally subnormal. He intended to call on local health and welfare authorities, through the bodies which represented them, to take a hand in mapping the joint future of the hospital and local authority services. One of the important qualities of local authority provision was the precision with which it ought to be able to cater for a whole gamut or graduation of different degrees of need. Mr. Powell doubted if now the availability of finance was the factor which limited provision. He intended to maintain something like the present ratio of local health and welfare authorities having half the capital expenditure on the hospital service, with, of course, increasing actual figures.

Integration

Prof. A. V. Neale's paper on 'Integration' ranged widely and eruditely, opening

with a historical survey. Since the second world war, he pointed out, there had been almost a global advance in psychiatric progress and research. Preventive psychiatry called for consideration of many possible predisposing factors—heredity, personal relationships, emotional, economic and physical stresses. There seemed no doubt about the desirability of giving special attention to preventive psychiatry in childhood.

One of the main objects of the new Mental Health Act was to remove rigid distinction between 'mental illness' and 'mental deficiency'. The word 'subnormality' expressed a clear positive natural disorder which should quickly remove any attitude of rejection or denigration of the individual involved. Trends indicated that the incidence of congenital and mental subnormality was not likely to diminish. Greater survival in infancy and childhood would incline towards some increase in numbers, at least in some areas. Hence the need to recognise, diagnose and classify as early as possible.

When Nature had created a state of arrested or incomplete development of the mind it was everybody's business to aim at the highest level of mental maintenance and every facility for self-respect and self-care. In the hospital for mentally subnormal there must be adequate and first-class clinical psychiatric practice and facilities for continued study of the patients in a medical, social and occupational sense. The general medical care must be good. The senior and junior medical staff, working as clinical teams, could accomplish a great deal. In general, the medical staff would have to be increased to remove the traditional common excessive overloading in numbers of patients under the care of one doctor.

More junior training centres for 'severely subnormal' children had been opened and Professor Neale hoped that all urban areas, at least, were providing for them in this way. Rural areas had geographical difficulties, and some boarding arrangements might have to be developed. Special care units could be suitably placed so that excessively difficult subnormal children were gradually made more amenable to life in a family: mother-like care from married women living in the vicinity of the special care unit was an excellent plan, especially if a child was not passed between too many people in the 24 hours.

Professor Neale spoke of the need to sharpen diagnostic methods. Wherever necessary, diagnostic assessment centres (best in relation to joint arrangements as between children's and general hospitals and nearby psychiatric hospitals) must be organised. In the future training of registrars and senior registrars for consultant appointments in the Mental Health Service he suggested that each should have experience in all branches on a rotating scheme, to include mental illness, juvenile psychosis, child guidance, mental subnormality, and special psychiatric units. This integrated training would have ultimate benefits for all and would abolish any existing or residual idea that the service for mental subnormality was something less than the other sections. The particular aspects of longitudinal care and training of the subnormal could not now be regarded as anything less than a most vital part of the mental health services. There was need for careful study of new policy in the staffing for mental subnormality.

Lastly, Professor Neale advocated that in the psychiatric hospital or group the medical advisory committee should have a wide membership—the consultant psychiatrists and S.H.M.O.s, consultant neurologist, consultant general physician, consultant paediatrician, consultant geriatrician, the medical officer of health (or his deputy) and some of the general medical practitioners in the area.

Treatment in Hospital

In a symposium of three papers on treatment in hospital, Dr. W. W. Sargant, physician in charge of the department of psychological medicine, St. Thomas' Hospital, testified to the effectiveness of a psychiatric outpatient and inpatient unit in a teaching hospital; Dr. Stanley Smith, Medical Superintendent of Lancaster Moor Hospital, Lancaster, described the Manchester region's policy of psychiatric units attached to general hospitals (or, in one case, a general hospital attached to a psychiatric unit); and Dr. W. A. Heaton-Ward, Medical Superintendent of Stoke Park Hospital, Bristol, dealt with the mentally subnormal aspect. This, Dr. Heaton-Ward pointed out, was concerned with the treatment of the accompaniments rather than the condition itself, and treatment was inseparable from training. He forecast the establishment in mentally subnormal hospitals of special units for subnormal patients with I.Q.s between 70 and 90. These should be self-contained as regards treatment and occupation, and would make great demands on staff as regards numbers and on the time of individual members. Dr. Heaton-Ward had a plea for M.P.s—to consider the long-term benefits to individual patients and to society of prolonged treatment in hospital rather than to try to find some legal loophole to prevent the compulsion without which such treatment might not be possible.

Reinforcing what Professor Neale had said about establishments, Dr. Heaton-Ward observed that the national average in the mental subnormality hospitals was one consultant to 700 patients. This ratio was deplorably low, but in some regions consultants had more than twice this number of patients under their care. It was difficult to understand how they could be personally responsible for the treatment of so many, as well as providing outpatient services. 'Unfortunately,' commented Dr. Heaton-Ward, 'we have not yet succeeded in convincing our colleagues in other branches of medicine, who influence regional hospital boards in fixing establishments, that we have progressed from a purely custodial approach to an actively therapeutic one in our care of the mentally subnormal.'

Community Care

When Prof. R. M. Titmuss came to consider community care he added the parenthesis: 'Fact or Fiction?' He said: 'It has been one of the more interesting characteristics of the English in recent years to employ idealistic terms to describe certain branches of public policy.' The practice had several unfortunate consequences. Public opinion (which included political opinion) might be misled or confused, and confusion was often the mother of complacency. The relentless London School of Economics dissection thereafter proceeded ruthlessly. The social legislation of 1946 and 1948, which gave local authorities practically all the legal powers they required to develop community care, had now been on the statute book for 13 years. In 1951 eight psychiatric social workers were employed full time by the 145 local health authorities. In 1959 there were 26. At this rate it would take until A.D. 2014 before someone could say that there was an average of one psychiatric social worker to each authority. Finance? Allowing for price changes, for the increase in our total population, for the larger increase in the number of mentally ill people seeking or needing treatment (judged by turnover, diagnostic and discharge rates), and for the increase in mentally subnormal people under statutory supervision and training, it was probable that a smaller amount per head was now being spent on community care for the mentally ill (as distinct from the mentally

subnormal) than in 1951. And what was being spent today was substantially less than the £4,900,000 paid out in compensation and expenses in dealing with fowl pest in Great Britain in 1959–60.

What the Minister had said about the reduction in the number of mental beds implied to Professor Titmuss quite a remarkable degree of optimism concerning the rapidly rising rate of readmissions; of faith in the capacity and willingness of general practitioners to participate in community care; of trust in the energy and vision of local health and welfare authorities; and of belief in the efficacy of the block grant as a means of developing community care. He wanted a specific earmarked grant to local authorities for community care services for the mentally ill and subnormal of £10 million for 1961–1962, central government grants for all social work students, training courses (irrespective of speciality) in the universities and technical colleges (courses to be established in 15 colleges by October 1962), and the appointment of a Royal Commission on the recruitment and training of doctors, with special reference to the need for education in social and psychological medicine. 'At present,' concluded Professor Titmuss, 'we are drifting into a situation in which, by shifting the emphasis from the institution to the community—a trend which in principle we all applaud—we are transferring the care of the mentally ill from trained staff to untrained or ill-equipped staff or no staff at all.'

The final session of the conference was devoted to public (which included professional) attitudes. Mrs. Mary Adams, the pioneer of documentary presentation on B.B.C. television, brought a series of recordings to illustrate such attitudes. These were as revealing as the statement of Miss Mary Grieve, the doctor's daughter who edits *Woman*. An article she had published on mental health invited readers to write to the magazine on any aspect of this subject. From the 10 million readers, experience showed that an article dealing with pregnancy would bring in thousands of letters, one on sexual maladjustment hundreds, and one on sterility a few hundred. The response to this article had been 14 letters, 4 of them from people engaged in the work. 'It suggests to me that a very great many of our readers, seeing the title, turned the page,' said the Editor of *Woman*.

LAURENCE DOPSON

What's to be done?

THIS is the fourth question circulated in this series, with the answers so far received. Further questions suitable for the series, and comments on the replies already published, will be welcomed.—*Editor.*

The Problem...

The child with hemiplegia seems to develop plantar flexion, from contracture of the calf muscles, despite regular physiotherapy by parents and hospitals from an early age. Does this imply:

(1) that more or more forcible physiotherapy should have been given;
(2) that physiotherapy makes no difference;
(3) that ankle-irons are nearly always needed; or
(4) that operation is often desirable before the age of 10 years?

...and some Replies

From A. J. INNES, F.R.C.S.
Orthopaedic surgeon to Stirling, Clackmannan and Perthshire Joint Hospital Board.

Many doctors who prescribe physiotherapy in cases of cerebral palsy have an uncomfortable feeling that it does not do much good. Yet they go on with it simply because they feel that something must be done and physiotherapy is one of the few possible lines of treatment. Apart from the really severe cases most spastics make some progress as they grow older. It is perhaps natural to give the credit for such improvement to any treatment that may have been prescribed. Certainly where a child appears to deteriorate in spite of physiotherapy there is no point in continuing with it and there is even less reason for making the treatment more strenuous.

Among spastics the children with hemiplegia have this advantage—that one side of their body is normal. This means that they are usually able to walk reasonably early. When such a child has a degree of equinus the important thing is to decide whether or not the deformity impairs function. Often it does not. Mild degrees of plantar flexion at the ankle are of little importance; and sometimes, indeed, even moderate degrees are quite useful in compensating for slight shortening of the affected limb. With more severe grades the foot will not fit the shoe and then something must be done to keep it at about a right angle. Where the deformity can be overcome by pressure, ankle-irons are indicated. These are rather clumsy but usually effective and often they can be discarded after a period of months or a year or two without recurrence of the deformity. Where fairly severe deformity is not controlled by irons an operation is required. It has been said that operation should not be done until growth is nearly complete. This is certainly not true of all cases, since good results have often been obtained in young children. In general,

however, the older the child the less scope there is for recurrence of the deformity. It is worth bearing in mind, of course, that taking cases of cerebral palsy of all ages the proportion requiring operation is not high and the number under ten years of age will be even smaller.

Briefly, then, my replies would be:

(1) No.
(2) Rather doubtful.
(3) No.
(4) No.

From W. S. TEGNER, F.R.C.P.
Physician in charge and director, Department of Physical Medicine, The London Hospital.

This question does not allow for a clear-cut answer, because every child is an individual and therefore needs individual assessment and treatment.

(1) It is probable that more physiotherapy should have been given, for we know that intensive personal care and enthusiasm on the part of the physiotherapist bears fruit. On the other hand, it is doubtful if focible physiotherapy is the answer to the problem if this distresses the child and causes fear and a lack of co-operation.

(2) Physiotherapy is of proven value. It is untrue to suggest that it is of no value. But the personality and devotion of the physiotherapist are of the greatest importance and this psychological aspect of treatment must never be overlooked or underrated.

(3) Again the question of leg-irons cannot be answered by a general statement. Many children can manage without them, but certain individuals are helped by them.

(4) Another difficult question to answer. If the medical advisers conclude that lengthening of the tendons is going to be necessary in an individual case, it is probable that this is best done before the age of ten years.

From K. BOBATH, M.D., D.P.M.
Consultant physician, Western Cerebral Palsy Centre, London

(1) Spastic contraction of the calf muscles is part of the extensor spasticity of the whole lower limb. Plantar flexion of the ankle with inversion of the foot results from the positive supporting reaction which produces spasticity not only of the calf muscles but also, to a lesser extent, of the proximal extensors. This means that spasticity of the calf muscles is not an isolated phenomenon, and physiotherapy that consists of forcibly stretching the calf muscles is therefore ineffective. It might, in fact, increase spasticity by repeatedly eliciting strong stretch reflexes with subsequent rebound contractions, and it fights a losing battle against the child's toe walking. More 'forcible' physiotherapy should not, therefore, be given, but physiotherapy which counteracts the spasticity of the whole side should be given as intensively as possible.

(2) Physiotherapy makes a difference, especially if given at an early age, if it:

(*a*) prevents the development of extensor spasticity and reduces it when already present; the child must take his full weight with the foot in dorsiflexion and the hip extended; the toes must not claw, the knee must remain mobile;

(*b*) reduces flexor spasticity in the arm and side flexors of the trunk, for this pattern of spastic contraction is associated with extensor spasticity of the leg; so long as a child 'hitches' his pelvis upwards to make a step he will not bend his knee or ankle;

(*c*) gives the child bilateral patterns, for instance, equal weight-bearing in sitting, kneeling and crawling on arms and legs; he must learn to feel his body not as two halves, using one for compensation and ignoring the other,

but as an entity; this will prevent the gradual increase of spasticity and also the possible contractures resulting from associated reactions through excessive use of the sound side;

(*d*) obtains normal equilibrium reactions of the affected side; the child will feel safer when weight-bearing on the affected lower limb while lifting the other; fear of falling is an important factor, causing an increase in spasticity.

(3) Ankle-irons are undesirable because they produce in time a permanent fixation of the ankle in a neutral position—that is to say, a contracture in a neutral position instead of in the equinus position. Though the former may be regarded as an advantage, it should be avoided if possible, since in many cases a pes cavus develops with a tightening of the plantar fascia. Furthermore, when fitting the child with an iron lying or sitting down, the heel may be in a proper position, but in standing, and more so in walking, spasticity increases and the heel may rise up in the boot unnoticed. Thus the child will still walk on his toes, though the heel of the boot is on the ground. In older children who walk and run on their toes all day long, *temporary* short plasters with dorsiflexed ankle and toes are recommended. After this, intensive physiotherapy on the above lines should be given, and if necessary a light night splint should be worn until a better walking and running pattern is established.

(4) Early treatment—i.e., treatment before the establishment of a contracture—should make subsequent operations unnecessary.

From Prof. J. CÍŽKOVÁ and V. VLACH. *University Children's Hospital, Prague, Czechoslovakia.*

(1) It is not clear whether most cases of infantile hemiplegia which have received good rehabilitation do develop irreversible contractures of the calf muscles and therefore irreversible plantar flexion. It is interesting to recall that many years ago, when rehabilitation was not so good as it is today, the results were less satisfactory. Not all cases of hemiplegia are spastic; we also see the paralytic form. Some cases develop a planovalga position of the feet. One can safely say, however, that the best way of preventing irreversible plantar flexion is by giving physiological rehabilitation from early childhood.

(2) The presence of persistent plantar flexion calls for intensive rehabilitation, though of course every case must be individually evaluated. The most important measure is passive and active exercise, together with massage and stimulation. We would like to draw attention to our successful use of pharmacological premedication before exercising. Hormone therapy (thyroid, testosterone) has also been used, together with very small doses of cortisone.

(3) We never use ankle-irons, because we feel that they have an adverse effect on the muscles.

(4) We operate as a last resort. Degree of function is the deciding factor; age is not important.

LETTERS TO THE EDITOR

Minimal Cerebral Palsy

SIR—During the course of my routine paediatric out-patient clinics and in particular the follow-up of all premature babies born in hospital in North Northamptonshire during the past seven years, I have become increasingly aware of the value of the concept of Minimal Cerebral Palsy in the early diagnosis of cerebral palsy and in the detection of children showing minimal signs of cerebral palsy. This is, of course, a term used in addition to the accepted classification of cerebral palsy, viz: the motor or physiological, the anatomical or topographical, and the aetiological systems.

By the term 'minimal cerebral palsy', I mean a minimal degree in the accepted signs of cerebral palsy by which such children are usually recognised and diagnosed. It does not necessarily follow that the child will be only slightly handicapped when viewed as a whole, although this is usually the case. Nevertheless, the failure to recognise and diagnose these children either soon enough or at all may be the most harmful part of the condition because of the mishandling and treatment they receive at the hands of their parents, their teachers and their fellow children and siblings.

It appears that the children diagnosed early and given simple but early physiotherapy have shown very considerable improvement in their spasticity (cerebral palsy) but this may be associated with maturation of the central nervous system in the children under 7, rather than treatment or perhaps because of both factors. As an example, I would quote a child (J.R.) who at 17 months had spastic legs with calf muscle and adductor spasm. At 5 years, he had only spasm in the calf muscles and far from scissoring, he had a wide-based gait with knee hyperextension to compensate for the calf spasm in standing and a slight forward lean of the trunk to obtain a better balance. There was no spasm in the hip adductors, the hip flexors or the hamstrings, only calf muscle spasm. And again, a child (G.P.) who had delay in her motor and adaptive norms of development to 15 months at a chronological age of 21 months and who walked on her toes at 2½ years of age. At 3 years she walked on her toes less but fell over rather frequently, especially if the ground surface she walked over was rough or slippery, and turned her right foot in more than the left, while at 6 years she only stumbled a little with her right foot, and only when she ran, and the spasm in her right calf was not easy to detect while that in her left calf was no longer detectable.

The older children picked up in routine out-patient clinics, I feel, could have been spared much trauma had they been diagnosed early. For example, the child who fell over backwards 'all in one piece' if a door banged or someone rushed past her at school unexpectedly, who had slight but definite calf muscle spasm increased considerably by emotional factors such as fear. The earlier diagnosis of 'epilepsy' and then 'hysteria' did nothing to lessen the child's emotionally increased minimal spasm nor appease the school-teacher father's intolerance of his thwarted ambitions for his child. And so one could go on quoting also 'clumsy' child after 'fidgety' child after 'backward' child after 'naughty' child for a very long time.

My ideas of minimal cerebral palsy can be defined under five headings:

(1) The very young infant, when the signs are minimal and difficult to elicit. There may be a feeding problem, a crying baby or a sleepy baby with paucity of movement. There may be persistance of tongue thrust. Granny may say that the baby does not 'hold right' in her arms when nursed; the hands may be clenched after three months of age. There may be an expressionless face. There is so often a significant obstetric and neonatal history.

(2) The minimal pyramidal cerebral palsy in infants, sometimes born post mature, more often premature and usually with a history of an abnormal pregnancy or labour and neonatal period. This would include the late sitters, late walkers, some of the stumblers, children who fall over more than they should and the clumsy child.

(3) The minimal athetoid cerebral palsy. Here one may mention those with late development of signs—perhaps up to three years or more. The fidgety child, the clumsy child, the child with the odd occasional writhing hand mannerism.

(4) The mentally defective brain-damaged child with minimal signs of cerebral palsy, such as clumsiness, dysdiadokinesis, astereognosis, atopognosis, minimal rigidity or spasticity, minimal athetosis.

(5) This is more speculative and is perhaps the main field for research. This is the child with minimal or no spasticity or athetosis, but marked disturbance of other functions of the brain now known to be frequently present in children showing obvious spasticity or athetosis. For example, marked disturbance of spatial perception and I am thinking of a child who was picked up, labelled mentally defective, but in fact had no concept of his own body image, nor could he draw any symbols at 9, but could be trained in this.

Other examples in this group are:

Disturbance of hearing: unilateral or bilateral; perhaps a high frequency loss as in kernicterics; auditory perception disturbance with regard to span, discrimination, analysis and synthesis of auditory stimuli—some of these are a developmental delay with a good prognosis. As examples, one could quote two children, who were premature and jaundiced, for whom E.S.N. schooling was being considered, but fortunately an assessment of hearing was made and audiometry showed considerable loss.

Visual perception disturbances: squints and disorders of eye movements.

Spatial perception disturbances, such as astereognosis with disturbed orientation in regard to structure, shape, size and body image.

Linked visuo-spatial perception defects with loss of figure background discrimination in pictures or in reality.

The marked psychological disturbances usually associated with cerebral palsy occurring in isolation from the spasticity and athetosis, e.g. behaviour problems, lack of drive, distractability, perseveration, fluctuation in performance. The hyperactive child.

Speech impairment, so-called congenital dysphasia, sometimes associated with a squint, many of these being familial and developmental. Possibly some stammerers.

Laterality complications: handedness and eyedness.

Linked visuo-spatial perception, visuo-motor perception and auditory-visuo-spatial perception disturbances causing disturbance of number concept, disturbance of writing ability and disturbance of reading ability.

I think the concept of Minimal Cerebral Palsy in the terms I have endeavoured to define has a place in aiding the early diagnosis of cerebral palsy and in the diagnosis of 'missed' cases and the atypical occult cases of cerebral palsy. Apart from paediatricians and neurologists, I think it could aid family doctors, school doctors, psychologists, E.N.T. surgeons, opthalmologists and paediatric psychiatrists. Perhaps teachers could be helped to see some of their children in terms of medical disability and not only in terms of educational problems, awkward children or behaviour problems?

How much impaired intellectual functioning in an educational environment can be correlated with this concept of minimal cerebral palsy is perhaps scope for more research and follow-up. It might seem reasonable to postulate that the neuronal death of neonatal hypoxia, etc., can cause minimal intellectual impairment as well as gross impairment and the importance of preventing minor degrees of hypoxia would then be seen to be greater.

The recognition and the acceptance of the concept of Minimal Cerebral Palsy would lead to a better recognition of the size of the problem and the challenge to society which cerebral palsy presents. Past estimates of numbers of cerebral palsied persons would be inaccurate because of the numbers of such patients who have previously been omitted in the various estimates made.—Yours, etc.

7 Cranford Hall, ROBERT WIGGLESWORTH
Kettering, Northants.

'Mongolism'

SIR—It has long been recognised that the terms 'mongolian idiocy', 'mongolism', 'mongoloid', etc., as applied to a specific type of mental deficiency have misleading connotations. The occurrence of this anomaly among Europeans and their descendents is not related to the segregation of genes derived from Asians; its appearance among members of Asian populations suggests such ambigious designations as, 'mongol Mongoloid'; and the increasing participation of Chinese and Japanese investigators in the study of the condition imposes on them the use of an embarrassing term. We urge, therefore, that the expressions which imply a racial aspect of the condition be no longer used.

Some of the signers of this letter are inclined to replace the term 'mongolism' by such designations as 'Langdon-Down anomaly', or 'Down's syndrome or anomaly' or 'congenital acromicria'. Several other signers believe that this is an appropriate time to introduce the term 'trisomy 21 anomaly' which would include cases of simple trisomy as well as translocations. It is hoped that agreement on a specific phrase will soon crystallise once the term 'mongolism' has been abandoned.—Yours, etc.

GORDON ALLEN (*Bethesda, Md.*)
C. E. BENDA (*Waverly, Mass.*)
J. A. BÖÖK (*Uppsala, Sweden*)
C. O. CARTER (*London*)
C. E. FORD (*Harwell*)
E. H. Y. CHU (*Oak Ridge, Tenn.*)
E. HANHART (*Ancona, Switzerland*)
GEORGE JERVIS (*Letchworth Village, New York*)
W. LANGDON-DOWN (*Normansfield, England*)
J. LEJEUNE (*Paris*)
HIDEO NISHIMURA (*Kyoto, Japan*)
J. OSTER (*Randers, Denmark*)
L. S. PENROSE (*London*)
P. E. POLANI (*London*)
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R. TURPIN (*Paris*)
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HERMAN YANNET (*Southberry, Conn.*)

Speech Therapy for Developmental Disorders

SIR—In his paper, '*Paediatric Aspects of Developmental Dysphasia, Dyslexia and Dysgraphia*' (*Cerebral Palsy Bulletin*, Vol. 2, No. 4, 1960, pp. 254-277), Dr. T. T. S. Ingram says it is very doubtful whether speech therapy has much place in the treatment of developmental speech disorders. He bases this conclusion on his observations on nearly 200 children referred to an Edinburgh speech clinic, suffering from what he terms 'specific developmental disorders of speech'. Rather more than half this number received speech therapy, while the others, living too far away, did not. No marked differences were observed in the rates of speech development in the two groups, he tells us, so he considers that maturation was the healer in both groups.

Nowhere in the article do we read of the nature of the observations on which the author based his conclusions. We are not told of the use of any standardised tests of articulation or language development. No ages are given for these children, nor is there any indication of the time elapsing between the original examination and the follow-ups. It is baffling to conceive how anyone compares two groups, each of about 100 children, with speech disorders so varied and complex, and describes them simply as 'similar'.

Carefully designed experiments have been reported from the U.S.A. which give ample evidence contrary to Dr. Ingram's conclusions. One, just published in the *Journal of Speech and Hearing Disorders* for February 1961, describes reliably administered tests of articulation applied to over 1,000 six-year-old children. Of these, 25 were selected for speech therapy and there were 25 matched controls. The difference between the scores achieved by the experimental group before and after 9 months' speech therapy were compared with the scores of the controls and submitted to statistical analysis. The result of the *t* test was significant at the 0.001 level—i.e. a *highly* significant difference in favour of those treated.

For over 12 years I have used a standardised test of articulation attainment to keep records of progress. Normally the child is tested at the first interview and retested at the end of each school term. The 'average' child with a speech defect (about 70 per cent of the total) shows rapid improvement in the first 3–4 months of treatment; then the rate of progress begins to slow up during the subsequent school terms. If the child is under 7 years and of average intelligence, this less marked improvement in the second term is usually observed whether speech therapy has been continued or not. Older and less intelligent children tend to relapse if treatment is not continued during the second term.

When comparing the progress charts of the untreated—i.e., those who attend only once every 3–6 months—there is seldom any evidence of rapid improvement in speech, but usually their graphs follow a line roughly parallel to that of the normal child's rate of acquisition of articulatory competence. Occasionally there is a sharp rise in the graph of the untreated, but I have yet to see a case in which there has been evidence of so rapid an improvement in any 3 months without speech therapy as the majority of the articulation cases show at the end of their first 3 months with speech therapy.

I can only conclude that Dr. Ingram's observations of cases must have been conducted at long intervals, perhaps 2 years or more, or that he was observing young children, possibly under 5. We have long been aware that practically all children (95 per cent) are speaking normally by 6½, although only about 50 per cent do so at 4½. But the point surely is that although some children improve quickly enough and young enough not to suffer from much confusion, others go through periods of real frustration through being unable

to communicate satisfactorily. The older speech-defective children grow, the more acute the problem becomes, because of the educational implications. There comes a time when, taking all the circumstances of age, intelligence, temperament, environment, etc., into account, the therapist decides that certain young children are ready for treatment that will shorten by many months their period of unintelligibility or being frequently misunderstood. To speed the relief from the effects of any handicap is surely a therapeutic aim. If the benefits of dentistry to young children were denied because their first teeth all fall out in the end, we could be condemning them to times of physical distress. Shall we similarly condemn the speech-handicapped child to times of mental distress?—Yours, etc.

Department of Speech Therapy,
The Churchill Hospital, Oxford.

C. E. RENFREW,
Chief Speech Therapist

BOOK REVIEWS

Psychiatry and the Public Health

By G. R. HARGREAVES

London: Oxford University Press, 1958, pp. 118, 12*s.* 6*d.*

Professor Hargreaves came to his university chair by an unusual route. Ignoring the conventional academic stairway he spent some time in industrial health and some in clinical psychiatry, including mental health work in the Forces, before taking charge of the Mental Health Section of the World Health Organisation. Only after several years of distinguished service in W.H.O. did he enter upon his present office at Leeds. International health work affects people in two principal ways. Some it raises to esoteric heights of abstruse specialisation, but others, faced with the task of helping the less-advanced countries to build up their services, come to grips with first principles. In this slim book, which reprints Professor Hargreaves's Heath Clark Lectures, we find a pointed and direct application of first principles to the orientation of psychiatry in a country which has for a long time tended to forget that first principles exist. The result is good reading for anyone interested in mental health.

By many accepted standards the author is a heretic, but his are wholesome heresies. He disagrees with those who describe psychiatry as a science, preferring to call it a practical art. How right he is! Medicine, as a late-comer in the scientific field, made the dangerous mistake of trying to be scientific so hard that it was in danger of forgetting its humanity. It has been said that any fool can practise purely scientific medicine and that many fools do; the wise physician knows that he is an artist as well as a technical specialist. This is even more true of psychiatry, but as the youngest infant in the medical family, psychiatry pretended to be a science to defend itself against the pretensions of its elder brothers and in the process almost convinced itself. As in medicine the would-be scientist was on the surest ground in pathology, so in psychiatry the emphasis has been on a pathological approach, and it is particularly useful at the present time to have a heretic who believes that the much-used term 'Mental Health' means something in itself and is not merely a synonym for the treatment of mental illness.

The classical instance of successful striving for health in a world of illness is probably to be found in the development of the British child health services. Faced with an enormous burden of morbidity these services first set about watching the child with the aim of detecting deviations from the normal at the earliest possible stage, when correction was fairly easy. In their next phase they watched the normal child and acted when they feared that deviations might be arising. Having found the factors which seemed to produce deviation, they advised the parents of normal children on the prevention of deviation and progressed logically to trying to influence the environment in the interests of normality or health. Hargreaves sees the road to mental health as something of this sort. There will be no true mental health so long as psychiatry is confined to the specialist treatment of the grossly mentally

sick, wherefore there must be an extension of psychiatric practice into fields where it can provide early diagnosis and treatment and advise on the prevention of substantial illness.

This, he feels, must involve at the very least some measure of psychiatric understanding in the general practitioner, the public health doctor and the health visitor. Much anticipatory action lies within their fields and they are admirably placed to detect incipient emotional difficulties and to recognise when the patient needs something which is beyond their scope. They are also in a position in which they can teach the art of healthy living, if they are willing to be teachers as well as healers.

Professor Hargreaves makes some pointed criticisms of medical administration in relation to psychiatry. With him we regret that administration has divorced the child-guidance clinic from adult psychiatry, but it must be recognised that the fact that child-guidance has grown up outside the hospitals, in the less pathology-minded environment of the education and school health services, has provided it with an opportunity of looking healthwards, an opportunity often well taken. Certainly his ideas of the child-guidance service of the future are well in line with the trends of progressive child-guidance work at the present day. If we interpret him rightly, he would like to see psychiatry divested of much of its present mystique and made an acceptable and accepted part of the general body of medicine. Certainly the mystique is responsible for much professional distrust of psychiatry and its practitioners, and after half a century of growth psychiatry ought to be able to stand on its own merits, with modesty as a much more becoming garment than mystery. That a professor of psychiatry should write in this tone—and, be it added, make his points with elegance and wit—is a major step towards the health of mental health.

JOHN D. KERSHAW

The Purpose and Practice of Medicine

Selections from Writings by SIR JAMES SPENCE

London: Oxford University Press, 1960, pp. 308, 42*s*.

This book contains 20 selections from Sir James Spence's bibliography of 53 publications. The articles are well chosen. It is with profit and great enjoyment that one reads these papers, which reflect so well the charm of their author, the personal, un-doctrinal way in which he tried to solve problems, the vast variety of his interests in the field of medicine, his broad-minded, humanitarian reflections on the medical profession, and his great common sense. Readers who do not know Sir James Spence's important achievements, and for whom he is only one of the famous names in the history of paediatrics in Europe, should carefully study these articles: *Nature of Disease; Children and Families; Care of Children in Hospitals;* and *The Need for Understanding the Individual.* It is most fortunate that the Editors have accomplished this collection of Professor Spence's papers, thus facilitating reference even to articles that were originally published in journals that are not easily accessible.

One reads with great pleasure the vivid picture of Sir James Spence by Sir John Charles in his introductory 25-page biography, in which are included some written souvenirs of Spence's friends. His

successor as Professor, Dr. Donald Court, calls him 'artist and scientist, romantic and realist, conservative and rebel, always a leader'. He was without doubt one of the great physicians of this century. He has contributed much to the advancement of our knowledge in paediatrics, especially its social implications. He was a rarely talented man, who used his own way in looking at problems and in trying to solve them, and in developing the science and art of being a physician.

This book should be included in every private and official paediatric library.

ARVID WALLGREN

The Natural Development of the Child

A Guide for Parents, Teachers, Students and Others

By AGATHA H. BOWLEY

Edinburgh and London: Livingstone, 1960, pp. 206, 10*s*. 6*d*.

This book conveys in quite a brilliant way, a living way, the normal stages of psychological development as seen by the psychologist. Real children are talked about—not statistical abstractions based on bogus tests. The norms have been well chosen, though one would have liked to have had normal limits rather than fixed milestones, and a discussion of the global view of the development one finds at any particular moment in a particular child. The references would be generous in a larger book, and are fairly up-to-date; they refer, on the whole, to other books rather than to journals. The educational needs of the child, in the broadest sense, emerge sanely, warmly, and with penetration.

The weakest parts of the book are those on psychodynamics and on psychological illnesses in the child—in other words, the old dichotomy between psychology and psychiatry goes on. The psychodynamics consist of a partial acceptance of one of the psychoanalytical views, which is only employed in some theoretical paragraphs, and not at all in the clinical examples. When it comes to treatment, the disturbed child is hardly seen as part of a pathological family—often the healthiest part! An incredibly superficial approach is suggested. Would that it worked!

This, then, is a book which succeeds as a Psychology of the Natural Development of the Normal Child. It is a bargain at 10*s*. 6*d*. It contains the sketchiest of psychiatry. Some of the more purely medical remarks are wrong. Could the next edition drop the section on *Children and the* (last) *War?* The only text for the next war is surely a religious one.

A. CROWCROFT

Special Education of Physically Handicapped Children in Western Europe

By W. W. TAYLOR AND I. W. TAYLOR

New York: International Society for the Welfare of Cripples, 1960, pp. 497, $3.50.

This is a fascinating title at a time when so many developments are taking place in a field where the educationist and the physician meet to help each other to a better understanding of the child's needs, where both have much to learn that can be applied in other spheres—in the education of the normal child; in the treatment

of other sick children; in the learning processes of the infant—but the reader will look in vain for an account of new work.

'Physically Handicapped' as here defined covers all types of handicap, including deafness and blindness and epilepsy, but not educational subnormality or emotional maladjustment. The first half of the book is devoted to a general survey in six chapters—*Organisation and Administration, Education of the Blind, the Deaf, the Orthopaedically Handicapped, Vocational Training, and the Education of Special Teachers.*

The second half has 27 chapters, one for each of the countries studied, Scotland, England and Wales, and Northern Ireland having a chapter apiece. In this half each chapter follows the same plan: history, definition of disabilities, services, teachers, organisation and administration. Of course the comparison of one country with another is most difficult—definitions vary, ages of admission to school and insistence on attendance are widely different—but there is no attempt to compare the incidence of disabilities and each country's success in providing education. There is a great deal to interest a reader in the account of what is done in each area but there is no over-all picture, no indication of what line of progress is most likely to be fruitful and what is already known to lead only to a dead end.

The confidence of the British reader must be shaken a little when he reads that all English teachers are paid by the Government, that retrolental fibroplasia occurred only in countries that provided incubators for premature babies, and that the day kindergarten for blind children in Vienna is unique—with no reference to the work in Sheffield or in Miss Freud's unit in London. There are some mysterious tables like those two about deafness in Scotland on page 401 that one feels must mean something if only there was a clue, and in the account of the work in England and Wales for the 'orthopaedically handicapped' the only reference to work for children with cerebral palsy is the opening of St. Margaret's School in 1947; neither the British Council for the Welfare of Spastics nor the National Spastics Society receive any reference. All children with tuberculosis are referred to as tubercular. There is no index, and the references given are almost entirely to Government publications, though the work of Miss Bowley and Professor Ewing are referred to in the English chapter.

Perhaps the subject is too great; certainly it needs more attention than it has received in this too superficial review, on which it is to be hoped that no international agency will feel disposed to make any special recommendations.

DENIS PIRRIE

Lecture Notes On Opthalmology

By PATRICK D. TREVOR-ROPER

Oxford: Blackwell, 1960, pp. 94, 12*s.* 6*d.*

Mr. Trevor-Roper's *Lecture Notes* are likely to supersede some of the other manuals which purport to help the beginner. Many of the so-called elementary books labour under two handicaps. First, they put in too much detail, as though an author were afraid to seem ignorant about subjects left unmentioned. Secondly, the presentation is fogged by lack of pictures, and especially coloured pictures. From these defects Mr. Trevor-Roper's book is refreshingly free. He has been able sympathetically to understand the difficulties of a final-year student striving to

grasp the essentials of opthalmology within a short time. Through the generosity of seven pharmaceutical firms, individually thanked in the Introduction, some of the 76 pictures are incorporated in excellent colour plates.

The author rightly stresses that pain and loss of vision are the cardinal symptoms of ocular disorder, and that there is a deep cleavage between the therapy of external and internal eye diseases. His first two chapters deal with the elementary anatomy and disorders of the external eye and its adnexa. Then follows a lucid account of squint, from which superfluous technical detail has been carefully pruned. Chapter 4 deals clearly with the painful red eye, a most important subject for the family doctor. The next two chapters are devoted to (*a*) gradual, and (*b*) sudden loss of sight in painless eyes free from external inflammatory signs; and here the neurological implications are not neglected. Chapter 7 on the injured eye contains much valuable guidance for the beginner, and a final chapter dealing with refractive errors is welcome. The average reader of these *Lecture Notes* will not intend to practise refraction. We should remember, on the other hand, that lay people are prone to believe ridiculous fallacies about eyesight. One of a family doctor's duties is to guard his patients against people seeking to exploit their ignorance concerning the scope and limitations of spectacles in dealing with refractive errors.

Mr. Trevor-Roper's *Lecture Notes* are well bound and printed on good paper. The price is low, considering the abundance and high quality of his illustrations, and the style of writing is attractive. The final-year students for whom it is intended should diligently apply themselves to this little book.

J. H. DOGGART

Human Growth

Symposia of the Society for the Study of Human Biology, Vol. 3

By J. M. TANNER

Oxford: Pergamon Press, 1960, pp. 120, 30*s*.

A symposium may be no more than a dreary re-statement of known fact, against a background obligato of thumped tubs, but this book is both refreshing and admonitory.

Part of its attraction lies in its wide scope. Professor Schultz discusses age changes in primates, Professor McKeown prenatal growth in man. Human growth is considered in relation to genetics by Dr. Tanner, to race and climate by Dr. Roberts, to nutrition and disease by Dr. Acheson and in its secular aspects by Dr. Boyne. Dr. Israelsohn's contribution is essentially a study in the use of data.

In his other writings Dr. Tanner has taken pains to point out what we do and do not know, how to find out and how not to find out, and one gets the same impression from this book, which seems edited rather than compiled. It becomes evident that in the study of human growth the amateur contributor, such as a paediatrician or M.O.H. who takes some measurements as part of his wider duties, must accept professional discipline, but this book will invigorate such amateurs rather than depress them. The paediatrician who considers 'teaching growth to students' a dull business will also get a fillip from the book.

Each paper has a good bibliography.

J. O. CRAIG

The Care of Invalid and Crippled Children

By A. WHITE FRANKLIN

London: Oxford University Press, 1960, pp. 158, 8*s.* 6*d.*

Published at the request of the Invalid Children's Aid Association, this book is designed to give almoners, health visitors, school nurses and social workers among disabled children an account of the medical conditions they will meet.

There are 16 short chapters by 11 acknowledged experts in their subjects. In addition to the common crippling diseases there are chapters on renal disease, diseases requiring dietary control, heart diseases, haemophilia and defects of vision, hearing and speech. The chapter on wetting and soiling, given balance by its editorial supplement, should be especially valuable.

It is encouraging to find the emotional reactions of the child and parent considered and a chapter is devoted to this aspect of hospital admission, but it is disappointing that the problem of helping the adolescent handicapped child to make a realistic adjustment is not discussed. Poliomyelitis is dealt with in just over a page, but little attention is given to the needs and management of the severely paralysed patient.

For its size this small book contains a great deal of information and will doubtless be of great service to those for whom it is intended.

F. E. JAMES

Cerebral Dominance and its Relation to Psychological Function Henderson Trust Lectures, No. 19

By PROF. O. L. ZANGWILL

London: Oliver and Boyd, 1960, pp. 48, 10*s.* 6*d.*

Professor Zangwill's excellent review of the literature on cerebral dominance, in his introduction and elsewhere, makes this book valuable as a reference work. The weakness in his presentation is to be found in his own data. Nevertheless, he establishes his main point, and does not overstep the bounds of his evidence, in holding that cerebral dominance is less clearly defined than was previously supposed.

His study is handicapped by the small number of cases (10) in his two series of left-handed patients with cerebral lesions of known location. Unfortunately he does not set out in clear detail how he selected these cases. He had three sets of data to compare: (1) the location, or at least the laterality, of the lesions; (2) the decision on the presumptive hemisphere of dominance; and (3) the presence or absence of dysphasia and also of dysgraphia and dyspraxia and the Gerstmann syndrome. Such qualitative characteristics are often difficult to evaluate, especially when they are mild or transient; and data of this kind cannot be accepted as confidently as measurements can. Thus a study of this sort is open to bias if the observer knows the laterality of the cerebral lesion at the time when he tries to assign the cases to the groups with one characteristic or another, such as the presence or absence of dysphasia. His position could be strengthened by setting out the conditions under which his patients were selected and assigned. This is very difficult to do with clinical material as it is usually presented to the investigator.

In the second section, on *Speech and Handedness*, there were 492 cases of

unilateral brain injury. There were 436 right-handed patients, of whom 105 had dysphasia. All the right-handed dysphasia cases had left hemisphere lesions; in none of these cases was the injury chiefly confined to the right hemisphere. There were 10 left-handed patients, 5 of whom had left hemisphere lesions and were dysphasic, while 5 had right hemisphere lesions, 2 of whom were dysphasic, one had agraphia, and the remainder were free of these impairments. The author states that the injuries in these cases were limited to one hemisphere, a statement which I think is too broad; for while a lesion can be quite circumscribed in the brain, the indirect effects of the lesion from pressure, circulatory and other changes could be quite diffuse. Diffuse disturbances of the cerebrum are frequently difficult to detect by present-day clinical means; and yet diffuse changes might be critical in the production of cerebral malfunctions, such as aphasia.

A second series of 10 left-handed patients with cerebral tumours of verified location was presented. Four of these cases had dysphasia associated with left hemisphere tumours, and one had dysphasia associated with a right hemisphere tumour. Minimal dysphasia was present in three other cases with left-sided and another with right-sided tumours.

A further series of 20 patients with specific educational disabilities (chiefly congenital word-blindness) were studied. It was found that 75 per cent of these people had members of their families who were left-handed; and also a high proportion of the cases in the series had other neurological manifestations.

The author concludes that the left-handed person is less likely to develop aphasia and allied disabilities from damage to the right hemisphere than is the right-handed person from damage to the left hemisphere. The left-handed person often develops aphasia from damage to the left hemisphere. However, the aphasia in the left-handed person is likely to be less severe and protracted than aphasia in right-handed people. In this regard left-handedness is advantageous; but the author feels that left-handed people are more susceptible to educational difficulties than right-handed people. He points out that cerebral dominance should be regarded as a graded characteristic in humans, there being degrees of left and right brainedness in individuals composing the population.

One is led to wonder whether, instead of being yoked together in one hemisphere, as has previously been thought, the functions of handedness and speech are actually independent of each other, though both are usually located in the left hemisphere.

Another question one is stimulated to ask on reading this monograph is: 'What correlation exists between handedness and aphasia, and to what extent do the pyramids decussate?'

Probably the most important practical conclusion to which Zangwill leads us is that lesions within the right hemisphere in left-handed patients can be attacked surgically with greater confidence that aphasia will not occur. We can also be more optimistic in the prognosis of speech recovery in left-handed patients with aphasia.

JOHN A. CHURCHILL

Abstracts

In collaboration with *Abstracts of World Medicine*, published by the British Medical Association, and with the kind assistance of the Excerpta Medica Foundation, *Courrier*, and *Obstetrical and Gynecological Survey*.

An Evaluation of the Treatment of Phenylketonuria with Diets Low in Phenylalanine

W. E. KNOX. *Pediatrics*, July, 1960, **26**, 1–11.

The influence of diets low in phenylalanine on the development of mental deficiency in patients with phenylketonuria is discussed in this paper from Harvard Medical School and the New England Deaconess Hospital, Boston. In a group of 466 untreated patients, the percentage with an I.Q. above 60 was 2.5. There was an everage fall of nearly 5 units in the I.Q. each 10 weeks that treatment was delayed. In 44 patients over 3 years of age at the beginning of treatment no dramatic changes in mental ability took place, but decreased frequency of seizures, restlessness, and irritability, with increased attentiveness, was noted. The response to treatment, which lasted from 3 to 61 months (mean 16.8 months), was studied in 43 children aged from 10 days to 36 months. In this group there were 18 times as many patients with an I.Q. above 60 and twice as many with a normal EEG as in the untreated group. These patients were then divided into those in whom treatment was started before and after 16 months of age. In the former, seizures were absent, the IQ was above 60 in 4 times as many patients as controls, and fewer patients had an abnormal EEG. Seizures occurred in 36 per cent of those in whom treatment was not begun until 16–36 months of age.

It is concluded that as progressive mental deterioration does not occur in the untreated patient, elevated plasma levels of phenylalanine cannot significantly injure the brain after the first few years of life. Treatment started in the early months of life is effective in the prevention of mental deficiency and neurological abnormalities. *G. de M. Rudolf*

A Syndrome of Phenylketonuria with Normal Intelligence and Behavior Disturbances

B. S. SUTHERLAND, H. K. BERRY, and H. C. SHIRKLEY. *Journal of Pediatrics*, Oct., 1960, 521–525.

The discovery of the association of phenylketonuria with mental retardation represented a considerable advance in medical knowledge. Recently, various authors have described patients with phenylketonuria but without mental retardation and in this paper from the University of Cincinnati, Ohio, 10 such cases are briefly summarised and to these the authors now add a further 2. One patient, a boy aged 3½, was mentally normal (I.Q. 93) and the other a girl aged 7, was slightly below normal (I.Q. 65). However, both children showed psychological disturbances manifested by a dull and expressionless facies, negative and apprehensive behaviour, emotional outbursts, and speech retardation. The boy was treated with a phenylalanine-restricted diet with resultant marked improvement in his behaviour. Although a similar diet prescribed for the girl was not maintained after her discharge from hospital she appeared to become 'livelier, more friendly, and co-operative'. The serum-phenylalanine level in both these children was considerably lower than that usually found

in those who are mentally retarded. A study of the published cases referred to above also revealed that lower serum-phenylalanine concentrations are often found in phenylketonuric patients with normal or near normal mentality.

David Morris

Chromosome Translocation as a Cause of Familial Mongolism

C. O. CARTER, J. L. HAMERTON, P. E. POLANI, A. GUNALP, and S. D. V. WELLER. *Lancet*, Sept. 24, 1960, **ii**, 678–680.

The hypothesis that chromosome translocation provides the mechanism by which some apparently normal parents have mongol children was tested in the case of a mongol child who had attended the Hospital for Sick Children, London, the chromosomal constitution of several of the child's relatives also being studied. The patient had 46 chromosomes, which included 2 representatives of chromosome 21, and an anomalous chromosome thought to be caused by a translocation between chromosomes 15 and 21. The patient's mother, maternal grandmother, and amaternal aunt each had 45 chromosomes, which included one chromosome 21 and also the 15/21 anomalous chromosome which occurred in the patient. The father and a brother of the patient each had 46 chromosomes which were apparently normal. Another brother and a cousin, who both died in infancy, had also been mongols. It appeared that the translocation 15/21 had occured at least as early as the embryogenesis of the grandmother.

H. Harris

The Clinical Picture of the Sturge-Weber Syndrome in Children. (*In Russian*)

R. B. SEJDINA. *Zurnal Nevropatologii i Psihiatrii*, 1960, **60**, 836–840.

The author describes 9 cases of the Sturge-Weber syndrome, recalling that the classic triad of signs in this condition consists of congenital angiomata of the face, congenital or early glaucoma, and angioma in one or other cerebral hemisphere. Areas of calcification of the brain may also occur, but these do not develop until after the third year of life, while other symptoms are convulsions, hemiparesis, and mental retardation. Of the 9 patients in this series 6 had angiomata of the face and scalp (in 4 cases also on the trunk and extremities), while all 9 suffered from hemiparesis, the angiomata being on the contralateral side. The paresis, however, had been present from birth in only one case, in 2 it followed a convulsion, and, in the other 6 it developed during the first or second year of life. Eight patients showed mental retardation. In one child, who died of sonne dysentery, the angioma in the right cerebral hemisphere was found at necropsy. In only one case was any abnormality of the optic fundus present, and this was on the contralateral side. Calcification of the brain was demonstrated radiologically in 3 cases, again on the opposite side—that is, the side of the cerebral angioma.

Inquiry showed that in 4 cases there was a history of a fall or a blow on the mother's abdomen during pregnancy; another mother had undergone an operation under anaesthesia in the 5th month of gestation; while another had been poisoned by swallowing some 'spirits' in the 2nd month. At birth 4 of the children had suffered from asphyxia, which may have had an effect in precipitating the disease; but, as the author remarks, it is difficult to distinguish the condition from birth trauma or acute encephalitis in young children, especially as the cerebral calcification and eye changes do not develop until later. One child aged 12 had no convulsions but suffered from severe attacks of migraine.

L. Firman-Edwards

Note: The recent monograph on this syndrome by Norman and Alexander (see

editorial on p. 212) should be consulted for fully up-to-date information on its diagnosis, pathology and treatment.—*A. V. Neale.*

Craniostenosis. (*In Russian*)
V. A. KOZYREV. *Zurnal Nevropatologii i Psihiatrii*, 1960, 1115–11119.

Craniostenosis as a nosological entity is comparatively rare, Günther having estimated its incidence at 1 per 1,000 births. The present report is based on the study of 63 cases referred to the Burdenko Neurosurgical Institute, Moscow, for operation between 1945 and 1958, 40 of the patients being male and 23 female (only 60 are considered in the discussion). The patients' ages ranged from 2 to 35 years. Craniostenosis was present at birth in 3 cases, and developed at varying periods after birth in the remainder. It has been attributed by some authorities to inflammatory processes in the meninges or cranial bones, and by others to birth trauma, rickets, endocrine disturbances or abnormal metabolism, while in some instances heredity has been invoked. The author considers that it is due to faults in the laying down and evolution of bone in the embryo.

The cases are classified according to the sutures involved. Mental retardation occurs earliest in patients in whom all the sutures are affected.

The cardinal signs are those of increased intracranial pressure (acute attacks of headache with vomiting), changes in the optic fundi (choked disks and secondary optic atrophy), exophthalmos (47 cases), psychological symptoms (18), epileptiform convulsions (15), meningeal symptoms (20), cranial nerve palsies, especially of the 6th nerve, and tonic or clonic nystagmus (16 cases). Craniograms showed absence of one or more sutures, and pneumoencephalograms revealed evidence of compression of the brain. The cerebrospinal fluid was usually normal. The results of operation were on the whole favourable; 34 children were able to attend school (only 8 had to go to special schools for poor sight), 7 went on to higher educational centres, while adults were able to resume work.

(No details of the operative procedure are given.) *L. Firman-Edwards*

The Surgical Treatment of Parkinsonism
F. J. GILLINGHAM, W. S. WATSON, A. A. DONALDSON, and J. A. L. NAUGHTON. *British Medical Journal*, Nov. 12, 1960, **ii**, 1395–1402.

The authors report from the University of Edinburgh the effect of producing electrocoagulation lesions in the globus pallidus, internal capsule and thalamus, separately or in combination, in an unselected group of 60 patients with Parkinsonism operated on during the 5 years 1955–60. All except one of the patients were under 65. The choice of site for the operative lesion was modified as experience accumulated. In the earlier cases the lesion was made in the globus pallidus, but in nearly a third of them an additional lesion was made in the ipsilateral thalamus to control tremor. In the second phase the lesion was sited in the thalamus initially, but rigidity was not resolved as completely as with a lesion in the globus pallidus. The present practice, which is to make an initial double lesion in the thalamus and globus pallidus, in each case close to the internal capsule, appears to produce the best results in controlling both tremor and rigidity.

In only one patient of the series have limb tremor and rigidity remained unimproved. Complications after the operation persisted in 6 patients who were successfully treated for tremor and/or rigidity, and several interesting temporary complications, such as transient dysphasia, were encountered. In one patient with moder-

ately severe diabetes, control was maintained with half the previous dosage of insulin after successful operation for Parkinsonism. There were no immediate postoperative deaths, but one patient died after many months of stupor resulting from a ventricular haemorrhage.

(It is perhaps a pity that in this interesting study there is no indication of the aetiological factors concerned in the pathogenesis of Parkinsonism in individual patients.) *J. B. Stanton*

Cataract in Children. Aetiological Aspect in Paediatric Material. (*In Swedish*)
A. BRONGE, B. HAGBERG, and L. MOLIN. *Nordisk Medicin*, Aug. 18, 1960, **64**, 1033–1038.

The authors discuss the aetiological factors in cataract of various types seen in 25 patients treated at the paediatric clinic of University Hospital, Uppsala, between 1938 and 1959. In 19 cases it was considered to be of pre-natal origin, 12 children having had a birth weight of less than 2,500 g. and 15 showing varying degrees of mental retardation, of whom 3 were mongoloid. A history of maternal rubella during pregnancy was obtained in 5 children, and 4 had cerebral palsy probably of pre-natal origin. A 'placental insufficiency' syndrome was diagnosed in 2 cases and was suspected in several others with dysmaturity. Other congenital anomalies were often present in addition to the cataract. Only one case of zonular cataract, occurring after probable rachitic spasmophilia, and 3 of diabetic cataract were seen. *G. von Bahr*

Viral Pneumonia of the Mother with Hemorrhagic Otitis in the Fetus
G. KELEMAN and J. H. NEAME. *Archives of Otolaryngology*, Aug., 1960, **72**, 163–169.

The possibility that maternal influenzal infection may be a cause of congenital deafness in the infant is discussed in this paper from Harvard Medical School and Massachusetts Eye and Ear Infirmary, with reference to the case of a woman aged 20 who died in the 23rd week of her first pregnancy after she had contracted pneumonia, in the course of which influenza-A virus (Asiatic strain) was isolated from her throat. Necropsy was performed on the foetus within 3 hours, but no virus was detected. However, among many other pathological signs, the temporal bones showed a haemorrhagic otitis of influenzal type.

The authors discuss the various factors involved, and give a warning that caution is needed in interpreting the findings.
F. W. Watkyn-Thomas

Hyperemesis Gravidarum as a Cause of Congenital Malformations. (*In German*)
R. HOHLBEIN. *Medizinische Klinik*, 1961, **56**, 93–95.

In the last 20 years various factors have been found to be potential causes of congenital malformations, including infectious diseases, disturbances of ovarian function, diabetes, vitamin and other food deficiencies, drugs, and disproportion between serum-albumin and serum-globulin levels. Here, attention is drawn to hyperemesis gravidarum as such a factor.

At the women's clinic of the University of Dresden, between 1949 and 1959, 120 children were born after their mothers had suffered from severe hyperemesis early in pregnancy. Of these children, 10 (8.3 per cent) had congenital malformations. In the same period, among 28,000 children born whose mothers did not have hyperemesis, 274 malformations were observed (0.98 per cent). This difference is impressive, and it seems probable that hyperemesis should be added to the list of factors which may cause congenital abnormalities.

It is clear from this observation that adequate treatment of hyperemesis is imperative. However, one should be

cautious about giving corticosteroids, since they themselves may also act as teratogenic factors. *Excerpta Medica abstract.*

Asian Influenza in Pregnancy and Congenital Defects

R. DOLL, A. B. HILL, and J. SAKULA. *British Journal of Preventive and Social Medicine*, Oct., 1960, **14**, 167–172.

Influenza due to the Asian strain of virus A first became epidemic in Great Britain in the latter half of 1957 and spread rapidly. An inquiry into the incidence of the disease in a general practice on the south-eastern outskirts of London showed that influenza was diagnosed in 28 per cent of 253 women of child-bearing age during the months of September and October, 1957. Accordingly 661 women attending the antenatal clinic at the Central Middlesex Hospital, London, between November, 1957, and March, 1958, were interviewed, and 240 (36.3 per cent) said that they had had influenza since the summer of 1957. Subsequently 175 of these women were delivered of 177 infants in the hospital; 128 of them were accepted (on the criteria given) as having had influenza and 47 were not. Of the 50 infants born to women with medical confirmation of influenza, 2 showed abnormalities, but in these cases the influenza had preceded conception by 2 months in one and 2½ weeks in the other. Of 22 infants born to women who had had influenza in the first trimester of pregnancy none had defects.

It is then pointed out that, taken at its worst, 63 of the 'accepted' mothers had influenza during the first trimester, and 2 of their infants had defects: on the other hand, of 66 women with 'accepted' influenza either before pregnancy or in the second or third trimester, 2 of their infants also had defects. An alternative comparison is between 2 defective children out of 89 born to mothers with influenza during pregnancy (2.2 per cent), and 27 such children out of 1,996 live births in the same hospital during 1953 (1.35 per cent). The authors conclude that there is no clear evidence from these data of congenital defects following Asian influenza in pregnancy, although a positive effect had been reported in earlier studies, and an increase in the stillbirth rate due to anencephaly in Scotland in 1958 and to a lesser extent in 1959 has been recorded, suggesting that Asian influenza in the early months of pregnancy could increase the risks of anencephaly in an area where its incidence is normally high. *F. T. H. Wood*

Causes of Death in Premature Infants. (*In English*)

A. SKOGRAND and K. HARNAES. *Acta pathologica et microbiologica Scandinavica*, 1960, **49**, 321–328.

Of 536 premature infants seen at the Children's Hospital, Oslo, during the 4-year period 1954–1957 a total of 120 died, the majority within 2 days of birth. Analysis of these 120 cases showed that the mortality was highest in infants with the lowest birth weight. Hyaline membrane was the commonest cause of death in the series (43 infants), and the commonest cause of death, usually in the first and second days of life, in infants weighing 1,000 to 2,000 g. at birth. Intracranial haemorrhage accounted for death in 22 infants, in 14 of whom the bleeding was intraventricular; other causes were atelectasis in 22 cases, pneumonia in 14, pulmonary haemorrhage in 9, and kernicterus, unassociated with Rh or ABO incompatibility, in 5. In the remaining 5, death was due respectively to oesophageal atresia, congenital cardiac defect, volvulus, hydrops foetalis, and intestinal perforations. (In general, these findings reflect the experience of most neonatal units.)

R. M. Todd

A Clinical and Experimental Study of the Effects of Exercise on Motor Weakness in Neurological Disease

J. A. R. LENMAN. *Journal of Neurology, Neurosurgery and Psychiatry*, Aug., 1959, **22**, 182–194.

Working at the Neurological Unit of the Northern General Hospital, Edinburgh, the author has studied the effects on motor weakness of isometric exercises, consisting in spring-resisted flexion or extension exercises of the biceps or triceps muscle carried out with the elbow flexed at 90 degrees. Each training session consisted of 30 maximal isometric contractions performed at 30-second or one-minute intervals. Changes in muscular strength were measured daily by means of a simple dynamometer giving direct readings, or electrically with a Wheatstonebridge circuit. Electromyographic recordings of the action potentials of the muscles were also obtained using surface electrodes. A few patients were given weight-lifting exercises to produce fatigue, each session lasting up to one hour. Five different groups were tested: (*a*) 6 healthy subjects; (*b*) 6 patients with muscular disorders; (*c*) 6 with disease of the motor neurone and one with syringomyelia; (*d*) 7 with demyelinating disease (disseminated sclerosis); and (*e*) 8 with miscellaneous diseases.

The results, which are discussed and illustrated by graphs and a table, were as follows. (*a*) The healthy subjects showed a gradual increase in muscular strength; (*b*) of the 6 cases of muscular disease, 5 showed an increase in strength, but in 4 the author suggests that this could have been due to other forms of treatment or to spontaneous recovery; (*c*) 2 of the 6 patients with motor neurone disease and the patient with syringomyelia showed significant improvement and no adverse effects were observed; (*d*) the 7 cases of disseminated sclerosis showed somewhat varied results; some improving and some deteriorating; (*e*) of the 8 patients with miscellaneous disorders 7 were improved (the diagnosis varied from poliomyelitis to protruding cervical disk). In some cases the improvement could have been attributed to other factors than exercise. In no case did fatigue have any harmful effects and it did not seem an essential factor in increasing strength.

It is concluded that as a means of increasing the strength of muscles paralysed by degenerative disease, exercise is probably without value, although it may prevent weakness from disuse. It is, however, worth while giving active exercises to patients who show evidence of recovery from paralytic lesions since this helps to increase the speed of recovery.

J. B. Millard

LETTERS TO THE EDITOR

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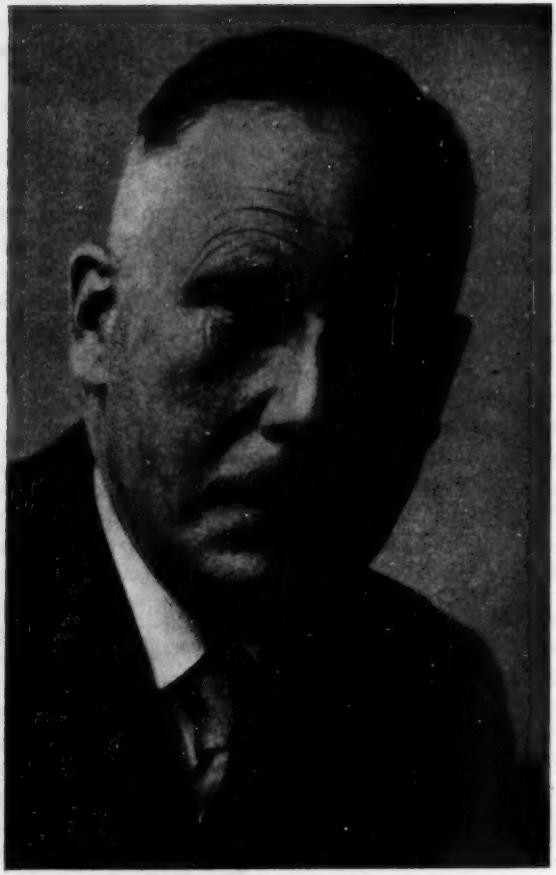

PROFESSOR B. S. ten BERGE

Born in 1896, Dr. ten Berge studied medicine at the University of Utrecht from 1914 to 1920 and then was resident in the Department of Obstetrics and Gynaecology of the University Hospital until 1923. He became president of the Department of Surgery at the Coolsingel Municipal Hospital in Rotterdam, and from 1926 to 1949 was head of the Department of Obstetrics and Gynaecology at that Hospital. From 1928 to 1949 he was also head of the Rotterdam Municipal Prenatal and Obstetric Service. From 1949 until his retirement in 1960 he was Professor of Obstetrics and Gynaecology at the State University of Groningen and director of the Obstetric and Gynaecological Department in the University Hospital.

Professor ten Berge deserves our special gratitude because by his example and by his own contributions he has increased our knowledge of the functioning of the placenta and because together with Professor J. Droogleevr Fortuyn he realised Dr. Heinz Prechtl's abilities and gave him the opportunities to work in the Obstetric Department at Groningen which are yielding a fine harvest. Dr. ten Berge has recently retired—but not to inactivity.

The Blood-Brain Barrier

THE 'blood-brain barrier' is an aspect of metabolic physiology which is a source of widespread misunderstanding and controversy, largely because of the implications of its unfortunate name. If it could be thought of instead as 'the blood-brain relationships of the central nervous system' many difficulties would be avoided. These relationships are clearly of the utmost importance, for, apart from their relevance to neuropharmacology, they have also been increasingly involved in the great advances of the last two decades in our understanding of the physiological biochemistry of the brain. Expression of current thought in these subjects often includes an almost mystical reference to the 'blood-brain barrier', which is held to account conveniently for any otherwise unexplained or unexpected observation. The result is such a mass of conflicting impressions that the subject has become almost disreputable.

For all practical purposes the idea of such a barrier was born in 1885 when EHRLICH[7] discovered that certain intravenously injected dyes (notably trypan blue) stained all tissues except the brain. For fifty years the 'barrier' was investigated with similar histologically identifiable agents, and an enormous literature accumulated. It is therefore not surprising that when the availability of isotopes made it possible to study *in vivo* brain metabolism, and when an apparent reluctance was demonstrated on the part of the brain to accept some of its constituents or their precursors from the blood-stream, this should be attributed to the same 'barrier' mechanism that had been shown to exclude trypan blue. Unfortunately as I, have recently pointed out in an extensive review of the subject[5], the dyestuff evidence has itself become discredited just when the neurochemists were using an extrapolation of it to account for some of their own observations.

Adding to the general confusion have been widespread misconceptions of the anatomical relations and physiological role of the cerebrospinal fluid, which have only recently begun to be resolved[12].

The dyestuff evidence in favour of a blood-brain barrier is of very restricted application because of the affinity these dyes have for plasma proteins, an association which might be expected to prevent their access to the brain in visible amounts. The virtual absence of cerebrospinal fluid protein accounts for the staining which happens after intracisternal administration of the dyes[11]. The important conclusion that the 'barrier' to trypan blue is all that can properly be investigated with this substance raises the question of how to explain the apparently similar limitation of access of ^{32}P and other materials of small molecular weight which play an active part in normal brain metabolism? Part of the answer lies in the recent discovery that many of the normal brain constituents are remarkably inert.

A substantial proportion of brain phospholipid phosphorus[2] and cholesterol[3] (presumably representing the myelin sheaths) would not be available for labelling in short-term experiments by circulating isotopes because it is inert. This means that, wherever the extent of isotopic labelling has been expressed (as is usual) as a concentration (radioactivity/total tissue content, or specific activity), the denominator has been 'diluted' by the inert mass, and the result has been an *apparent* retardation of entry, much of which is not real. This 'dilution' effect of inert constituents achieves all the greater prominence when the brain is compared with other tissues (e.g. liver) in which the same substance may be wholly labile. Furthermore, the larger uptake by the developing brain is easily accounted for since this is the time at which incremental growth of the adult inert compartments is taking place. The extent to which the hindrance to entry of metabolic substances is only apparent, and can be accounted for in this way, can only be determined when the absolute sizes of the metabolically inert compartments in each case have been measured, a task which has scarcely begun. At present, however, it may well be misleading for neurochemists to think in terms of a blood-brain barrier to a metabolite until they have demonstrated that the rate of the metabolite's entry places a restriction on its metabolism. Much confusion will be avoided if, until it is proved otherwise, the rate of passage of a metabolite from blood to brain be considered a *reflection*[5] of its metabolism rather than a limiting factor in it. The entry of glucose into the brain is very rapid, a fact consistant with its being the organ's chief oxidisable substrate; while the entry of cholesterol into adult brain is as slow as might have been predicted from its predominantly inert role.

The blood-brain barrier cannot be discussed without mentioning current controversy over the existence or otherwise of an extracellular space in the central nervous system. EDSTRÖM[6] has pointed out that much of the slowness of entry of certain substances from the blood might be accounted for if there were no extracellular space and everything had to cross several cell membranes in order to penetrate the tissue. DAVSON[4] has countered this suggestion by claiming to have demonstrated that 'there *is* an appreciable extracellular space, entry to which is impeded by a blood-brain barrier.' Electron microscopists, however, are remarkably unanimous in declaring that there is no extracellular space to be seen in the central nervous system. This is so even in experimental oedema, when the increased water content of the tissue is seen to be selectively confined to the interior of one type of glial cell[8,9]. There is no agreed solution to this problem at present, but it cannot be denied that the absence of an extracellular space could well account for most of the observations. The only serious obstacle to acceptance of the idea is a healthy reluctance to believe that brain should differ in this way from all the other tissues of the body. But such reluctance does not constitute evidence, and if we could be convinced by biophysicists and others that it would be possible for the brain to

acquire its nutriment at the observed fast rate entirely through cells, the case against such a space would be greatly strengthened. The idea would also have to be reconciled with current views on the mechanism of nerve impulse conduction.

The subject of the blood-brain relationships for pharmacological agents is even more complex. A demonstrable, partial dependence for tissue penetration on lipid solubility and degree of ionic dissociation[1] should not obscure the influence of those metabolic processes on which the drug acts. Such physical characteristics as lipid solubility influence penetration into other tissues as well as brain and may not therefore contribute much to the idea of a blood-brain barrier. It should also be recognised that arrival in the brain in pharmacologically recognisable form and in therapeutically effective quantities is not necessarily synonymous with blood-brain penetration, although the first two are, of course, the only useful parameters in practical therapeutics. Rapid metabolism or alteration of a drug on entering the brain could render it ineffective as well as unrecognisable by specific assay techniques and give a spurious impression of failure to penetrate.

Kernicterus is another topic which concerns the blood-brain barrier. Current views almost invariably attribute the brain damage in this condition to the presence of large circulating amounts of unconjugated bilirubin, and the evidence for this view is at first sight impressive. No attempt is made, however, to account for the entry of the (protein-bound) bilirubin into the brain, and, even if this intellectual difficulty is overcome, why should it enter such circumscribed regions? It is no longer believed that there are differences between adult and neonatal brains in their relationships to blood-borne dyes[10]. The older hypothesis was that focal damage occurred first with a hypoxic pattern of lesions, and that these areas were subsequently and quite gratuitously stained. Recent findings indicate that 'indirect' bilirubin dissolves easily in brain lipids *in vitro*, that it interferes with cell metabolism, that kernicterus occurs at lower bilirubin levels in the presence of certain protein-uncoupling drugs, and that it appears to be averted by prophylactic replacement transfusion. But none of these findings explain the peculiarly selective entry. Bilirubin, like other protein-bound substances, will invariably enter previously damaged areas of brain, whether the brain is adult or foetal and whether the pigment is conjugated or not. Both varieties are firmly attached to protein, and there seems no reason to distinguish between them and protein-bound dyestuffs whose non-specific entry into brain lesions of whatever kind is fully documented. Furthermore, the pattern of the stained areas bears a remarkable resemblance to the distribution of those parts of the brain which are differentially susceptible to hypoxia. It does seem, therefore, that the old 'primary damage' hypothesis may have been prematurely dropped in favour of a hypothesis that bilirubin is the cause of the damage, which still remains to be proved in terms of the known distribution of lesions and the known blood-brain relationships for protein-

bound substances. So far there is nothing—not even the demonstrable relationship between kernicterus indidence and levels of serum bilirubin—which necessarily implicates the bile pigment as the cause of the brain damage; and, the more sophisticated our views on hypoxia become, the less does this seem to be excluded as a primary factor in kernicterus.

The blood-tissue relationships of the central nervous system are so complex as to make it even more desirable than usual to avoid misunderstandings and misconceptions. It seems at least possible that clear thinking has been hindered more than helped by the concept of a 'barrier' in the usual meaning of the word.

JOHN DOBBING

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Down's Disease?

THE condition known as 'mongolism' in Western Europe or 'Down's disease' in Eastern Europe is unlikely to be a new disease. It probably accounted for many of the cases described by SEGUIN as a 'furfuraceous' form of cretinism and has until recent years often been confused with that disease. We owe to LANGDON DOWN[5] in 1866 the first adequate description of the syndrome. Since that time, it has had an established place in the text-books. However, not all contemporary text-book accounts are correct in all particulars and there is considerable variation between individual patients. There may be *formes frustes* of the disease and mosaics may occur, but in the main the condition is clear-cut and may be recognised with fair confidence at birth, though mistakes do occur.[9] Characteristic changes are seen in many parts of the body. Of these, the pattern of the dermal ridges has been most intensively studied and lends itself to quantification. As Dr. SARAH HOLT points out in her article in this *Bulletin*, expert study of palm-prints alone is of high diagnostic value. As with other forms of mental retardation, more attention was drawn to the condition as living conditions improved, the infantile death-rate fell and compulsory education was introduced. In recent years there seems to have been a considerable increase in the cases surviving, though there is no evidence of an increase in the incidence at birth. Information on this point will accumulate as more studies of the newborn are published.

The term 'mongolism' was used because of a facial resemblance to true Mongols. Objectively the resemblance is very limited and there is no difficulty in distinguishing cases of 'Down's disease' in countries with a Mongolian population. For this reason among others the use of the term 'mongol' to denote a disease state is unfortunate and the description 'a mongol Mongol' might give rise to confusion. It is true that some features are common to the anthropological and pathological groupings. Of these, the epicanthic folds are the most readily identified, but the plica marginalis, the horizontal fold seen in the upper eyelid of many Mongolians, is not a special feature of the pathological condition as it occurs in West Europeans; nor are prominent cheekbones. Limited nasal development was probably another feature suggesting the use of the current term.

It is interesting to speculate on whether there is any possible common factor in the determination of pathological 'mongolism' and the peculiarities of true Mongols, such as the epicanthus, but it is clear that the clinical syndrome occurs in all or most racial groupings and that there is no justification other than tradition for the use of the present term. Even in 1925, when CROOKSHANK wrote *The Mongol in our Midst*, there

was enough scientific information available to discredit his fanciful views regarding an anthropological basis for the classification of mental defect. In retrospect, JULES VERNE has proved to be a much more realistic writer.

'Mongolism' was first described in Europe because conditions there were favourable for its early recognition. There is no good evidence as to the relative incidence in different geographical areas, but this seems to be roughly the same in different parts of Europe and North America.[8] It was maintained until recently[3] that 'mongolism' did not occur in African negroes—the cases described in the United States were attributed to white admixture—but LUDER and MUSOKI[7] have shown that the condition is in fact not uncommon in African negroes. Similarly in Japan, though the syndrome received little attention before the Second World War, recent studies initiated into the effects of atomic explosions have disclosed a considerable incidence in areas unaffected by irradiation. It seems likely that the number of surviving cases in underdeveloped countries will increase as the infantile death-rate falls.

The discovery by LEJEUNE and others that 'mongolism' is due to a chromosome anomaly offers the possibility of revision of the nomenclature. Various new names have been suggested,[1] based on the type of anomaly. There are exceptions to the general rule of 21 trisomatism, so any description based on this anomaly would have to allow for other possibilities, such as translocation, and those that may be discovered subsequently. Some of the suggested terms are perhaps too cumbersome to catch on for general usage—thus, it is doubtful whether 'congenital acromicria' or 'trisomy 21 anomaly' will have a general appeal.

Since the term 'Down's disease' is already well-established in many countries, it would be simplest if this were adopted as the usual generic term. As the system of nomenclature of chromosomes and their anomalies becomes standardised, it will be possible, in particular cases where minute accuracy is desired, to qualify the generic term with agreed code numbers —e.g., 'a case of Down's disease (3×21)', or 'Down's disease ($15 + 21$)'. This should eliminate any suggestion of racial prejudice or misunderstanding which is inherent in the current Western term.

BRIAN H. KIRMAN

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The Choreiform Syndrome and other Work done by Heinz Prechtl

On June 14, Dr. HEINZ PRECHTL from Groningen, The Netherlands, was a guest of the Association for Child Psychology and Psychiatry at the Royal Society of Medicine, where he spoke on the work he has been doing over the past five years. He started by discussing the choreiform syndrome, which he has seen in numerous children who were first referred to hospital because of behavioural difficulties at school. When doing EEG readings on them, Dr. PRECHTL observed that, although their EEG pattern was normal, there were occasional bursts of activity from the muscles of the neck and temporal region. He then extended his observations by doing polymyographic studies and found bursts of unexplained activity in recordings from practically all the muscles. On palpating the muscles he could feel 'flicks' of activity. These children find it difficult to concentrate, are restless, and are often described by their mothers as very active children who cannot keep still. With undiscerning teachers they may run into a lot of trouble.

Dr. PRECHTL went on to outline some of his work on the newborn. In particular he discussed the follow-up work done on infants who were abnormal in the neonatal period, as judged by the neurological examination which he and his colleagues have elaborated in Groningen. He has demonstrated some well-marked correlations between the findings in the neonatal and follow-up periods. Dr. JAN DIJKSTRA[1], who has been responsible for the follow-up of the children between the ages of 1½ and 4 years, has been particularly impressed by the children described in the newborn period as having the hyperexcitability syndrome, for a statistically significant proportion of them go on to develop the choreiform syndrome described above. This syndrome, Dr. PRECHTL explained, is not seen before the age of 3 years.

Studies of the normal population in the area by Dr. PRECHTL and his colleagues have demonstrated the choreiform syndrome in 20 per cent of the boys and 9 per cent of the girls, making an overall incidence of about 14 per cent. These figures were confirmed by several independent observers.

This was only one of the several startling findings, supported by statistical data, which Dr. PRECHTL put forward. He was keenly questioned by members of the Association, for whom some of Dr. PRECHTL's ideas call for a reorientation of views on the origins of behavioural problems in childhood. It seems likely that 'minimal brain damage' contributes to far more of these problems than has hitherto been

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supposed. It may be also that some of the 'clumsy' children, about whom there is much discussion at the moment, owe their disabilities to minimal neonatal brain damage.

Dr. PRECHTL will shortly be contributing an article to the *Bulletin* on the choreiform syndrome. Some account of his other studies will be found in the Proceedings of the Second (1960) International Study Group on Child Neurology and Cerebral Palsy, which is being published as a *Little Club Clinic in Developmental Medicine* this Autumn.[2]

MARTIN BAX

2. See also Mac Keith, R. C., 'Prognostic value of the neurological examination of the newborn', Editorial, *Ibid.*, 1960, 2, 189.

NOTICE

Children and Newborn Infants in Hospital

THE American Academy of Pediatrics[1] has recently issued a booklet, *The Care of Children in Hospital*, which will interest all those concerned with the welfare of children or the planning of children's wards. The emphasis is on the total care of children in paediatric units which are part of a general hospital. This trend away from special children's hospitals, with a complete orientation towards the needs of children, is becoming a necessity in view of the complexity of modern medicine and surgery. It increases the burden on the paediatrician, who must ensure that there exists throughout the staffing of the general hospital an awareness of the special needs of children and adolescents. These needs are both physical and emotional and it is no easy matter to reconcile maximum safety and physical care with the equally important need for avoidance of emotional trauma.

The American Academy's recommendations are reasonable and sensible if rather unexciting. The views expressed are in line with modern attitudes toward such problems as cross-infection, the need for a play and educational area and the desirability of free visiting. Human relationship are discussed and the need for proper education of the medical and nursing staff is given due consideration. Lip service is given to the idea that Mother may in some circumstances be admitted with her child but in this respect the proposals fall short of what we in Britain might have wished.

This is a helpful and authoritative booklet; armed with it and our own Platt Committee Report the Paediatrician need not go naked into the committee-room when he makes demands for improvements in paediatric accommodation or services. If the paediatrician has also care of newborn infants he will be wise to obtain a copy of the American Academy's companion booklet, *Hospital Care of Newborn Infants*, which gives similar information on the planning and procedures recommended for the newborn and premature nurseries. However, this booklet which was last revised in 1957, gives little guidance on human relationships or the necessity for proper training of personnel in the handling of mother-baby relationships.

Thomas Oppé

[1] American Academy of Pediatrics. 1801, Hinman Avenue, Evanston, Illinois, U.S.A.

The Parents Rôle in the Child's Development

THIS topic, which was chosen for the World Child Welfare Congress in Brussels in July 1958, is so vast as to be beyond the range of any one congress. This is still more true if its delegates come, as they did there, from many parts of the world. The representation in Brussels was so international that, if the emphasis had been placed on cultural differences in regard to the parents' rôle, many interesting comparative data might have emerged. As it was, the subject matter was mainly subdivided according to the children's age levels, with one section devoted to *The Home* and another to *Community Development Projects and the Family*. It was therefore not surprising that a specialised subject like *The Handicapped Child* provided the main theme for discussion by only four speakers.*

The most interesting address, at any rate from the child psychiatrist's point of view, was given by Dr. Dell' Acqua, who said:

'Our concept of rehabilitation is always based on knowledge of the "person" hidden behind the handicap. It is only by knowing the "person" or the character of the particular child that we can work towards sure integration, since it is the individual psychological structure of each handicapped person which explains certain of his reactions to his handicap'.

From this statement, taken out of its context, it might be inferred that Dell' Acqua's approach to the assessment of an individual's 'psychological structure' was somewhat facile. However, in fact he set out to show that this 'structure' needs to be understood not so much in terms of the child's innate psychological structure as of the parents' reaction to the child and his handicap. Dell' Acqua was mainly concerned with the consequences of adverse parental attitudes. Experience had taught him that the crux of the problem of likely success or failure in *social* integration often hinges on the nature and quality of parental attitudes.

No one doubts that there are parents who have no real love for their children. Child psychiatric experience teaches us that, in a country which supports extensive medical and social services, the physically defective child who is unloved and unwanted by its parents may present a less difficult problem than the perfectly normal child in a similar parental plight. Indeed, some of the most damaged yet physically unimpaired children are those whose parents consciously deny their hostility or who deliberately hide it in other ways. Among the more subtle of these parental rôles is a kind of oversolicitude that makes the child feel so doubtful about his independent capacities for self-management that he becomes cripplingly inhibited. The adverse motivations of a solicitude which presents itself as a loving preoccupation with potential harm (to the child) may be even more misleading when lavished on a physically handicapped child. Sometimes the guilt from which such parents also often suffer in regard to a handi-

* 'Problems arising for the parents of the handicapped child', by E. Kong (Switzerland); 'Help to parents of handicapped children', by A. Parloor (Belgium); 'Relations between parents and school in the case of the physically handicapped child', by J. Mengal (Belgium); and 'The social and professional integration of handicapped young people', by U. Dell' Acqua (Italy).

capped child may have even more confusing outcomes. The normal children in such a family are not only neglected, apparently in the service of the physically defective child, but may be forced into disliking it and envying its 'privileges of helplessness'.

One of the most surprising findings in child *psychiatric* practice is how often a patient's organic handicap or other disability turns out to fit into—i.e., to 'satisfy'—a kind of predestined Fate pattern of conflict in one or both parents. It is a tragic 'satisfaction' which, if it can be brought into focus and usefully understood by the parent(s), may bring about encouraging changes in the whole family atmosphere.

A remarkable example of such a predestined 'satisfaction' was in a boy of superior intelligence who developed a school phobia on winning a place at a grammar school. At the age of 6 his mother had taken him to another child-guidance clinic because of his difficult behaviour. When the mother now came for psychiatric advice she carried another child in her arms. This was an idiot girl of 4 years, who was also a severe athetotic. The mother, a highly disturbed woman, made it clear that her life and interests were dedicated to this uncomprehending creature which she carried or wheeled about almost unceasingly. She had refused repeated offers to have the child admitted to an institution. Fortunately we had taken the precaution of communicating with the previous clinic and had excellent case-notes at our disposal.

The astonishing feature which we were to learn was that this woman used to wheel her active 6-year-old son about everywhere, including to school. In the other clinic's opinion she had a compulsion to deprive him of his normal capacities in order to keep him in a permanent state of helpless infancy. The advent of a helpless idiot child, therefore, did no more than fulfil her peculiar pre-requirements.

The most unusual feature of this unusual case is that her pathological pseudo-solicitude was documented elsewhere 6 years in advance.

A pre-school or grossly physically handicapped child is necessarily extremely dependent for its life experiences, and for its understanding of them, on its parents. To a professional 'outsider' it seems possible that in some physically handicapped cases, in which progress is bafflingly disappointing, therapeutic efforts should be concentrated on the parents rather than on the child. It is in this context that the congress *Proceedings* proved especially interesting. Only in countries with full employment, where high standards of care are freely available from the State, can pronouncements be made on their effectiveness in terms of parental responsibility. It was therefore interesting to note that the only contribution of its kind was that of Mr. D. Jones of Great Britain, on 'Families who do not or cannot respond to the general community services at their disposal'. However, as Dell' Acqua underlined, responsiveness is as much a question of feeling as of will.

AUGUSTA BONNARD

Palm-Prints

IT has long been believed that a man's external appearances must reveal something of his internal nature. In the 18th century, when LAVATER made his celebrated studies of physiognomy in what he thought was a scientific fashion, he believed he had defined facial characteristics so that it was possible to reach definite conclusions about a person's character by merely studying his face. He was tempted to go further, and he related the extraordinary skin, covered with deer-coloured hair, that he found in a girl to an argument her mother had had with a neighbour about a stag while she was pregnant. In his essays such curious beliefs and fancies are intermingled with acute observations. Despite its obvious limitations, the study of the face can clearly reveal something of the owner's nature, as witness the Parkinsonian facies.

Other external parts of the body can yield almost equally valuable information. The hand has always attracted attention and, while the traditional gypsy still tells your fortune for the traditional reward, research workers are now busy submitting palm-prints to scientific investigation. However, the gypsy bases her fortune-telling on the flexion creases in the hand, whereas it is the dermal ridges whose study from a medical point of view is discussed by Dr. SARAH B. HOLT, of the Galton Laboratory, on another page. It is not always realised that these ridges, rather than the skin-creases, are the subject of modern dermatoglyphic study.

Various ancient peoples seem to have used finger-prints as a means of identification, and Biblical sources are sometimes quoted in support of this view. In the Book of Job (*Ch.* 37, *v.* 7), for instance, we find:

> '*He sealeth up the hand of every man; that all men may know his work.*'

These prints have certainly been used as signatures since early times.

The modern story probably starts with an English engraver—THOMAS BEWICK—who was so struck with the beauty of finger-prints that he made engravings on wood of his own prints and used them as marks of identification on some of his works. BEWICK died in 1828, and shortly before that, in 1823, JOHANNES PURKINJE, the Bohemian physiologist, first classified the dermal ridge patterns in a thesis presented to the University of Breslau. But the real father of modern finger-prints was Sir WILLIAM HERSCHEL (not the astronomer), who first began to use finger-prints for identification purposes around 1857 in Bengal, to prevent the impersonation which was then highly prevalent in the Bengal courts. The experiment was so successful that the Indians came to regard finger-prints with superstitious awe. A system of classifying finger-prints was later devised by Sir EDWARD HENRY, at one time private secretary to

the governor of Bengal, and in 1901 he established the finger-print bureau at Scotland Yard. The impetus for the bureau came from a Home Office committee's report of the previous year. This committee had also heard evidence from Sir FRANCIS GALTON, who, with the help of data supplied by HERSCHEL, had demonstrated that, unlike other external physical characters, the details of the ridges on the fingers and palm persist unchanged throughout life. The genetic features of individual finger-prints and palm-prints are now being studied at the Galton Laboratory.

MARTIN BAX

Prayer for a Deformed Girl

Almost murdered in her mother's womb
And saved too soon for an easy way,
Poor Head-on-Shoulder (she wears no name
But that from me) has limped along this day
To fill my thoughts with mimic shaping
As though she were myself.

She has humped away, but yet crooks here.

Her shape I lived in as many griefs
As take to mouth my aping care—
From the mourning of her back-hand birth
To this day that brought her hunching here,
Linked by the years that call up silence
Where tears are never told.

But I feel her weep, and pray for her.

This night shall form my wishes, for
As dreams grow huge and shadow-deep,
I shall use the dark that sorrows share
To hang her shame and shape in sleep
And make her straight with the irons of love
Until the morning comes.

God please that this I may do for her.

Jack Marriott, 1961.

ORIGINAL ARTICLES

The Influence of the Placenta on Cerebral Injuries

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A CONNECTION between diplegic cerebral palsy, birth asphyxia and prematurity was established about a hundred years ago by Little (1862). Since then many authors have confirmed that asphyxia at birth may cause serious neurological and mental disturbances. Animal experiments have also shown that changes in behaviour may follow asphyxia at birth, and anatomical abnormalities have been observed in these animals.

However, no direct correlation between the severity of cerebral aberrations and the degree of hypoxia induced can be established. Attention has been chiefly centred on the events occurring during parturition, whether terminated artificially or not, and most investigations have been retrospective. It has been universally agreed that complications at birth can result in severe asphyxia with injuries to the central nervous system, but these injuries are also found after normal deliveries. Opinions differ as to whether oxygenation of the baby is diminished by normal delivery. Eastman (1954) maintains that this is so, but Walker (1954) was unable to confirm it.

Asphyxia can be due to various causes, including prolonged contraction of the uterus, umbilical cord abnormalities, narcosis, diminished maternal respiration and cardiac defects, as well as instrumental delivery. The consequences of asphyxia depend on its origin and the duration of apnoea, but chiefly on the oxygen reserve available to the child during labour. If the foetus has suffered from latent oxygen lack during the last few weeks *in utero* the consequences of asphyxia at birth will be more severe, and injuries to the central nervous system can be anticipated sooner. The risks of cerebral palsy occurring in association with prematurity has been demonstrated by Eastman (1954). In a retrospective investigation he compared 500 cerebral palsy babies with 500 healthy babies. Of the babies with abnormalities 39 weighed under 1,600 grammes, whereas only 2 of the controls weighed less than this figure.

The question remains whether prematurity in itself was the cause of these neurological abnormalities, or whether the existing hypoxia resulted in premature birth, oxygenation during pregnancy being significant as well as events taking place during labour.

At the Obstetric Clinic of the University of Groningen, Dr. H. F. R. Prechtl carried out a neurological investigation from 1954 onwards of all neonates born following pre- or para-natal complications. Dr. J. F. Dijkstra (1960) then made prospective follow-up examinations of the infants. The following complications were considered to be indications for neurological examination of the infant:

1. Haemorrhage during pregnancy.

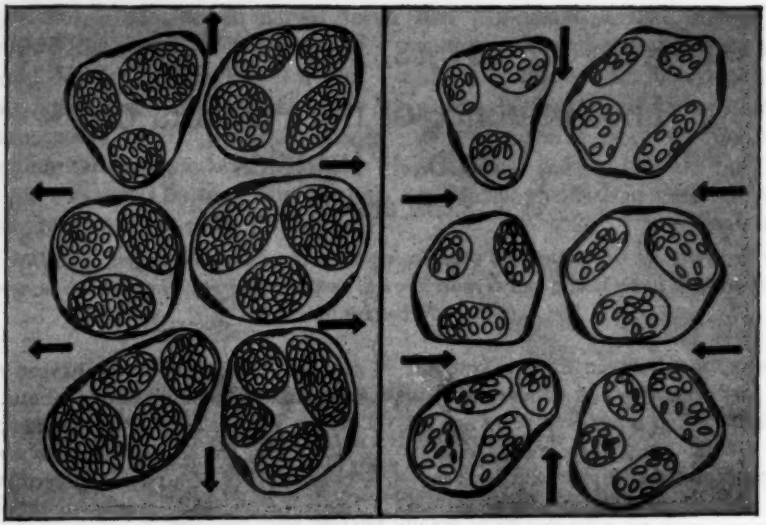

Fig. 1.
Influence of filling and emptying of the capillaries in the villi on the circulation of blood in the intervillous spaces.

2. Moderate to severe toxaemia.
3. Hypertension.
4. Slow or irregular heart sounds.
5. Breech presentation.
6. Version and extraction.
7. Caesarian section.
8. Forceps delivery.
9. Apnoea or post-partum asphyxia.

Results

Neurological abnormalities were found in 38 out of 60 children who had presented symptoms of neonatal hypoxia. There had been the possibility of hypoxia, toxaemia, fluxus, prolonged labour, etc., in 41 babies, of whom 19 showed neurological abnormalities (6 cases of Caesarian section for toxaemia, *4* neurologically abnormal; 11 forceps deliveries, *7* abnormal; 10 spontaneous head presentations, *5* abnormal; 1 version and extraction, *1* abnormal; 3 breech deliveries, *2* abnormal). In 18 infants there was no reason to suppose that there had been any hypoxia; only 3 of these presented neurological aberrations in the neonatal period (10 breech presentations, *2* abnormal; 3 Caesarean sections, *no* neurological abnormality; 5 forceps deliveries, *1* abnormal).

From a follow up 1½ to 4 years later, hypoxia appeared to be the main cause of permanent neurological disturbances. These were present in 48 per cent of infants who had suffered from neonatal hypoxia against 11 per cent who had not.

These findings throw doubt on current obstetric principles. As obstetricians we have realised that if we wish to improve our prenatal care, great attention must be paid to the possibility of hypoxia in the child in pregnancy and throughout labour. Even more interest must be directed to the anatomy and function of the placenta, and especially to the foetal circulation.

According to our research, circulation in the placental bed is maintained by the capillary pulse in the villi (ten Berge 1955a). The placenta is rich in pre-capillaries, making up the paravascular network (Bøe